

## ONCOLOGY KNOWLEDGE HUB · FOR HEALTH PROFESSIONALS

# Cutaneous Lymphomas

## A mini review for clinicians

For postgraduate residents (dermatology, medical, radiation and paediatric oncology, haematology, pathology, nuclear medicine, medicine), practising clinicians and medical students

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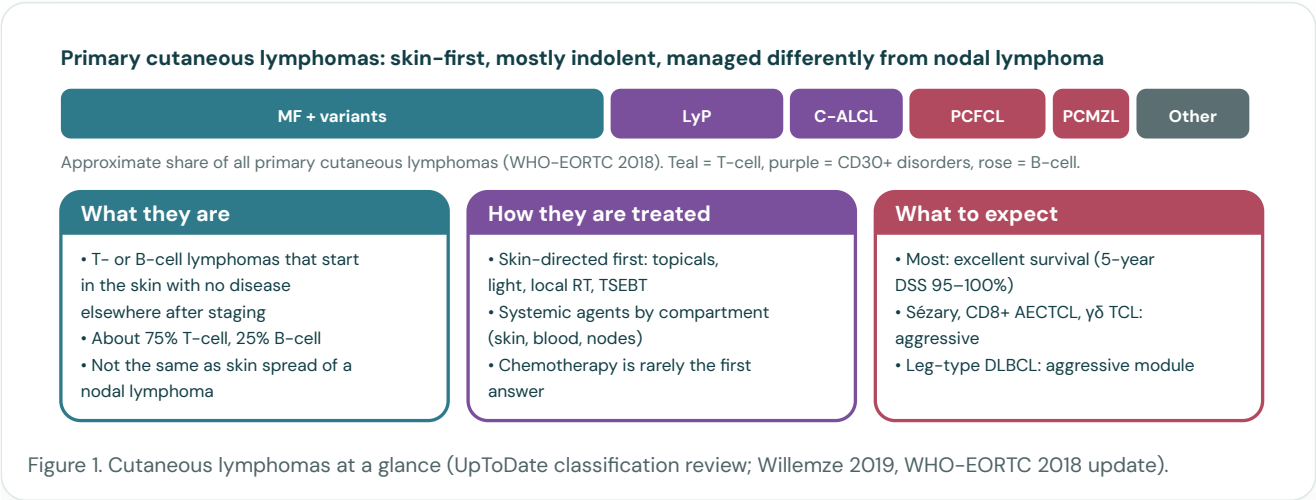
Updated 17 October 2026. Educational summary written in our own words from current guidelines and primary trials; no drug doses; not advice for an individual patient. Verify current guidelines and Indian regulatory approvals before treatment decisions. Patients and families: please see the patient guides at drharitha.co.in.

# Cutaneous Lymphomas

Haematological cancers, part 5 · Mycosis fungoides and variants, Sézary syndrome, lymphomatoid papulosis and cutaneous ALCL, SPTCL and rare T-cell types, follicle centre and marginal zone B-cell lymphomas · Skin-directed treatment · Compartment-based systemic therapy · Key points for clinicians · Last updated 17 October 2026

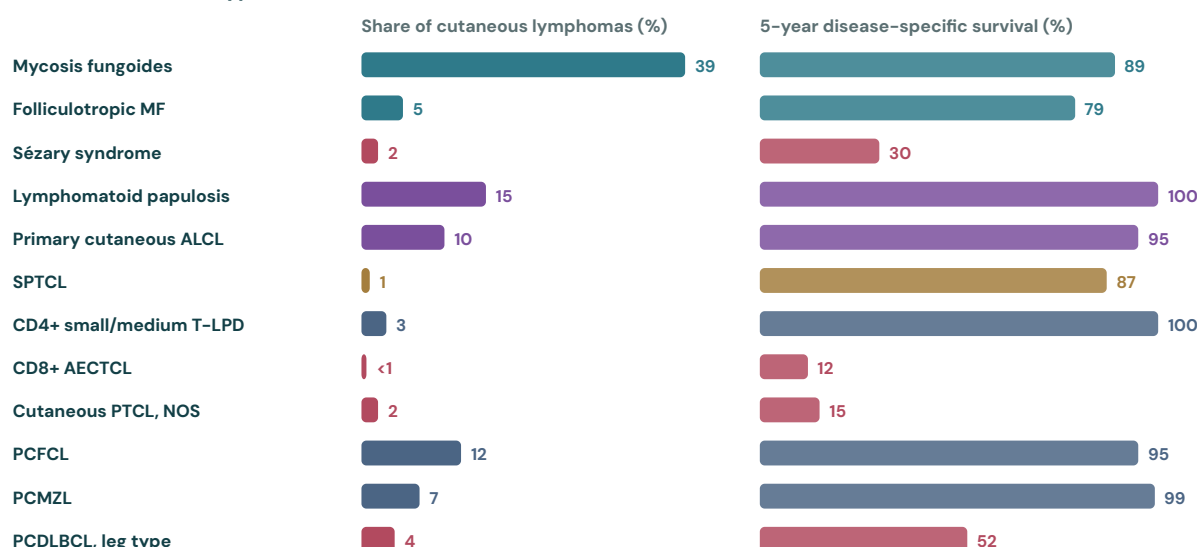
## 01 At a glance

- **Primary cutaneous lymphomas start in the skin** with no disease elsewhere after staging. They behave differently from nodal lymphomas with skin spread, and most are indolent: treating them like systemic lymphoma causes harm.
- **About 75% are T-cell, 25% B-cell.** Mycosis fungoides (MF, about 40% of all) and the CD30+ disorders (lymphomatoid papulosis and primary cutaneous ALCL, about 25%) make up about 90% of cutaneous T-cell lymphomas; follicle centre and marginal zone lymphomas are most B-cell cases.
- **Diagnosis is clinicopathologic:** early MF looks like eczema or psoriasis for years; biopsies (≥4 mm, off topicals, often repeated) are scored with the ISCL/EORTC 4-point system; a T-cell clone alone proves nothing.
- **Stage MF and Sézary syndrome by TNMB** (skin, nodes, viscera, blood by flow cytometry); other cutaneous lymphomas by their own TNM and a PET-CT to exclude systemic disease.
- **Treatment follows stage and compartment:** early MF → skin-directed (topical steroid, mechlorethamine, NBUBV/PUVA, local RT, TSEBT); tumours and LCT → brentuximab vedotin; erythroderma and blood → ECP or mogamulizumab; allogeneic HCT is the only cure for advanced MF/SS.
- **Indolent B-cell and CD30+ disorders:** low-dose RT, excision, topical or intralesional steroid; lymphomatoid papulosis is often just observed; multiagent chemotherapy is rarely right.
- **India:** hypopigmented MF (children, young adults, darker skin) is mistaken for leprosy, vitiligo or pityriasis alba; in one South Indian series ATLL was 17% of cutaneous T-cell lymphomas, so HTLV-1 testing has a place here too.



## 02 Clinical presentation, common traps & causes

### How common each type is, and how it behaves



Indolent (teal, purple, navy) far outnumber aggressive (rose). Sézary syndrome, CD8+ AECTCL, γδ TCL and leg-type DLBCL behave like systemic lymphomas.

Figure 2. Frequency and 5-year DSS by type (WHO–EORTC 2018 update, Willemze 2019, as tabulated in the UpToDate classification review; <1 shown as <1).

### Who gets it, and why

- **Mycosis fungoides:** about 6 per million per year in Europe and the USA (about 4% of NHL); peak age 55–60, men twice as often as women, more common in Black populations; also seen under 35 with the same course. Not inherited.
- **How MF shows:** itchy patches in the "bathing trunk" area (limited in about 30%, generalised patch/plaque in 35%), tumours in about 20%, erythroderma in 15%; also poikiloderma, alopecia, follicular papules, keratosis pilaris-like lesions. Nodes are involved in about 30%; viscera are rare without tumours or erythroderma.
- **Variants:** folliculotropic MF (the commonest variant in adults; early-stage form behaves like early MF, tumour-stage form like tumour MF); pagetoid reticulosis (solitary acral plaque, benign); granulomatous slack skin (rare; second lymphoma risk, often Hodgkin); unilesional MF (curable locally). **Hypopigmented MF** is the commonest variant in children and darker skin (33% of MF in a Southeast Asian cohort); often CD8+; responds well.
- **Sézary syndrome:** older adults; rapid erythroderma with nodes, severe itch, alopecia and palm/sole keratoderma; median survival 3–4 years.
- **Lymphomatoid papulosis:** 1.2–1.9 per million per year; peak in the fifth decade but any age; crops of papules that necrose and heal in 3–8 weeks; lifelong risk of a second lymphoma.
- **SPTCL:** median age 36 (20% under 20), women twice as often; up to 20% have autoimmune disease (lupus, juvenile arthritis, Sjögren, type 1 diabetes); painless fat nodules on legs and arms.
- **Cutaneous B-cell lymphomas:** PCFCL (median age 51, slight male excess) on scalp, head and trunk; PCMZL in adult men on trunk and arms, linked with tattoo pigment, tick bites and (in Europe) Borrelia.

- **India:** in a 6-year South Indian series, MF was 57.1% of cutaneous T-cell lymphomas, ATLL 17.1%, PTCL-NOS 17.1% and ALCL 2.9%, with poor outcomes in ATLL ([Khader 2016<sup>\[1\]</sup>](#)). At KEM Mumbai, hypopigmented MF was found in 0.17 per 100 skin biopsies, mostly in young men, about 3.8 years after onset, and mimicked Hansen's disease ([KEM series<sup>\[2\]</sup>](#)).

### Common traps

- **Years of "eczema", "psoriasis" or "fungal infection" on the buttocks and trunk.** Persistent patches in sun-protected skin that change size and shape: biopsy, after stopping topicals.
- **Pale patches treated as leprosy, vitiligo, tinea versicolor or pityriasis alba.** Hypopigmented MF needs a biopsy, sometimes several.
- **Erythroderma called "drug rash" or atopic dermatitis without blood flow cytometry.** Sézary syndrome needs flow cytometry and a skin-matched clone.
- **A clonal T-cell result read as lymphoma.** Clones occur in benign dermatoses, drug eruptions and older people; pseudolymphomas (actinic reticuloid, lymphomatoid drug or contact reactions) can look identical under the microscope.
- **LyP called a high-grade lymphoma and given CHOP.** Large CD30+ atypical cells in a self-healing papule are LyP; multiagent chemotherapy is inappropriate.
- **Fat nodules called panniculitis or lupus profundus for months.** SPTCL needs a deep biopsy that includes fat; lupus profundus is painful and has no clonal TCR.
- **ALK-positive CD30+ skin nodules** are usually systemic ALCL in the skin: stage fully.

### SHARE WITH PATIENTS: SYMPTOMS, SWELLINGS AND DIAGNOSIS

Plain-language myth pages written for patients and families. Send them from the clinic to reinforce what you discussed.

[Itching all over the body is always just a skin problem?](#)

[Lymphoma can't occur outside the lymph nodes?](#)

[All lymphomas are the same cancer?](#)

[Lymphoma is contagious and can spread between family members?](#)

[Lymphoma occurs only in elderly people?](#)

## 03 Diagnosis, workup & staging essentials

### Making the diagnosis

- **Biopsy technique:** stop all topicals except emollients for 2–4 weeks; punch of at least 4 mm from the most indurated lesion; several biopsies, sometimes over time. Excisional or deep incisional biopsies (with fat) for nodules, SPTCL and B-cell lymphomas; shave biopsies are inadequate. LyP: excise two or more fresh, non-necrotic lesions.
- **MF:** small to medium cerebriform cells along the basal layer and in the epidermis (epidermotropism) without spongiosis; Pautrier microabscesses are specific but present in about 38%; haloed lymphocytes are the most robust clue. Typical phenotype CD2+, CD3+, CD4+, CD5+, CD7–, CD8–; hypopigmented MF is often CD8+. Large cell transformation (LCT): large cells >25% of the infiltrate or forming nodules.
- **ISCL/EORTC score for early MF** (Figure): clinical up to 2, histology up to 2, clonal TCR 1, immunopathology 1; 4 points makes the diagnosis ([Pimpinelli 2005](#) [3]).
- **Sézary syndrome** needs all three: diagnostic (or compatible) skin biopsy; erythroderma over 80% of body surface; B2 blood ( $\geq 1000/\mu\text{L}$  CD4+CD7– or CD4+CD26– cells) with a TCR clone matching the skin ([Olsen 2022](#) [4]).
- **CD30+ disorders:** LyP shows a wedge-shaped mixed infiltrate with large CD30+ cells (type A in about 75%); primary cutaneous ALCL has sheets of large cells with CD30 in more than 75%; both are usually ALK–negative (nuclear ALK → systemic ALCL).
- **SPTCL:** atypical cells rimming individual fat cells; CD3+, CD8+,  $\beta\text{F1+}$ , cytotoxic granules; CD4–, CD56–, EBER–.
- **$\gamma\delta$  T-cell lymphoma:** TCR $\gamma\delta$ +, CD56+, usually CD4–CD8–, reaches epidermis and ulcerates.
- **B-cell:** PCFCL is BCL6+, BCL2– (mostly), MUM1–, not graded; PCMZL is BCL2+, BCL6–, CD10– with monotypic cytoplasmic Ig in plasma cells. Strong BCL2, MUM1, FOXP1 and IgM point to leg-type DLBCL.

#### Early mycosis fungoides: the ISCL/EORTC 4-point score

| Clinical                                                                                                                                                                                                                                                                                                                      | 2 points max | Histology                                                                                                                                                                                               | 2 points max | Molecular                                                                                                                     | 1 point | Immunopathology                                                                                                                                         | 1 point |
|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|--------------|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|--------------|-------------------------------------------------------------------------------------------------------------------------------|---------|---------------------------------------------------------------------------------------------------------------------------------------------------------|---------|
| <ul style="list-style-type: none"> <li>• Persistent or progressive patches/thin plaques, plus:</li> <li>• non-sun-exposed site</li> <li>• variation in size/shape</li> <li>• poikiloderma</li> <li>• 2 extras = 2 pts; 1 = 1 pt</li> </ul>                                                                                    |              | <ul style="list-style-type: none"> <li>• Superficial lymphoid infiltrate, plus:</li> <li>• epidermotropism without spongiosis</li> <li>• lymphoid atypia</li> <li>• both = 2 pts; one = 1 pt</li> </ul> |              | <ul style="list-style-type: none"> <li>• Clonal TCR gene rearrangement</li> <li>• Supports; never diagnostic alone</li> </ul> |         | <ul style="list-style-type: none"> <li>• &lt;50% of T cells CD2, CD3 or CD5+</li> <li>• &lt;10% CD7+</li> <li>• Epidermal/dermal discordance</li> </ul> |         |
| Total of 4 or more points = mycosis fungoides                                                                                                                                                                                                                                                                                 |              |                                                                                                                                                                                                         |              |                                                                                                                               |         |                                                                                                                                                         |         |
| Biopsy rules: stop topicals (except emollients) 2–4 weeks before; punch of at least 4 mm from the most indurated lesion; often several biopsies over time. Pautrier microabscesses are specific but present in only about 38%; haloed lymphocytes are the most robust single clue. HTLV-1 serology if from an endemic region. |              |                                                                                                                                                                                                         |              |                                                                                                                               |         |                                                                                                                                                         |         |

Figure 3. Diagnosing early MF (Pimpinelli 2005; UpToDate MF diagnosis review).

**The cutaneous lymphoma key: where the cells sit and what they express**

|                     |                                                          |                                                  |
|---------------------|----------------------------------------------------------|--------------------------------------------------|
| <b>MF / Sézary</b>  | Epidermis (epidermotropism); CD3+ CD4+, CD7 or CD26 lost | Blood: CD4+CD7- or CD4+CD26- by flow; TCR clone  |
| <b>LyP / C-ALCL</b> | Dermis; large CD30+ cells (C-ALCL: >75% of cells)        | ALK negative; ALK+ → look for systemic ALCL      |
| <b>SPTCL</b>        | Fat lobules; rims adipocytes; CD8+, βF1+, cytotoxic      | CD4-, CD56-, EBER-; HAVCR2 germline; HLH         |
| <b>γδ TCL</b>       | Epidermis to fat, ulcers; TCRγδ+, CD56+, CD4-CD8-        | Aggressive; median survival about 15 months      |
| <b>CD8+ AECTCL</b>  | Epidermis, necrotic; CD8+, CD4-, CD5-, cytotoxic         | CD2, CD7 variable; median survival under 2 years |
| <b>PCFCL</b>        | Dermis, spares epidermis; BCL6+, BCL2-, MUM1-            | Head/trunk; t(14;18) 0–40%                       |
| <b>PCMZL</b>        | Nodular, Grenz zone; BCL2+, BCL6-, CD10-                 | Monotypic cytoplasmic Ig in plasma cells         |
| <b>PCDLBCL, leg</b> | Sheets of large cells; BCL2, MUM1, FOXP1, IgM strong     | MYD88 L265P about 60%; aggressive module         |

**Clinicopathologic correlation decides: a clonal T cell, CD30+ cells or atypia alone do not make a lymphoma (pseudolymphomas, drug eruptions, LyP mimics).**

Figure 4. Cutaneous lymphoma key (UpToDate MF, SPTCL, LyP, rare subtype, PCFCL and PCMZL reviews; simplified).

**Before treatment**

| Area             | What to do                                                                                                       | Why                                              |
|------------------|------------------------------------------------------------------------------------------------------------------|--------------------------------------------------|
| Skin             | Whole-skin examination; % body surface by lesion type (the patient's palm ≈ 0.5%); photographs                   | T stage; response baseline                       |
| Nodes and organs | Examination and imaging; excise clinically abnormal nodes (FNA is not enough)                                    | N1–N3; Nx if not biopsied                        |
| Blood            | CBC, chemistry, LDH; flow cytometry (CD4/CD7/CD26, TRBC1) and TCR clone in MF/SS                                 | BO–B2; CLIPI                                     |
| Viruses          | HTLV-1 where relevant; HBV and HCV before methotrexate                                                           | ATLL; safe treatment                             |
| Imaging          | MF: CT or PET-CT for T2b, T3, T4, nodes >1.5 cm, LCT or B2 (else chest X-ray). Other cutaneous lymphomas: PET-CT | M and N stage; primary vs secondary skin disease |
| Marrow           | Only for unexplained cytopenias or equivocal imaging; core biopsy                                                | M1a                                              |

Staging mycosis fungoides and Sézary syndrome: TNMB, then the stage

|       |                               | Clinical stage |                   |
|-------|-------------------------------|----------------|-------------------|
| T1    | <10% BSA patches/plaques      | IA             | T1 NO MO BO–1     |
| T2    | ≥10% BSA patches/plaques      | IB             | T2 NO MO BO–1     |
| T3    | One or more tumours ≥1 cm     | IIA            | T1–2 N1–2 MO BO–1 |
| T4    | Erythema ≥80% BSA             | IIB            | T3 NO–2 MO BO–1   |
| NO–N2 | Nodes normal or not effaced   | IIIA           | T4 NO–2 MO BO     |
| N3    | Node effaced (Dutch 3–4)      | IIIB           | T4 NO–2 MO B1     |
| M1    | Visceral (M1a marrow only)    | IVA1           | Any T NO–2 MO B2  |
| BO    | <250/μL abnormal CD4 cells    | IVA2           | Any T N3 MO BO–2  |
| B1    | Between BO and B2             | IVB            | Any T any N M1    |
| B2    | ≥1000/μL + skin-matched clone |                |                   |

Early = IA–IIA (skin-directed treatment); advanced = IIB–IV. Image (CT or PET-CT) only for T2b, T3, T4, nodes >1.5 cm, large cell transformation or B2. Biopsy enlarged nodes by excision (FNA is not enough). Marrow only for unexplained cytopenias. Other cutaneous lymphomas use a separate TNM (T1 solitary, T2 regional, T3 generalised).

Figure 5. MF/SS staging (Olsen 2007 and 2022; Kim 2007 for non-MF cutaneous lymphomas; UpToDate staging review).

SHARE WITH PATIENTS: SCANS, BIOPSIES AND STAGE

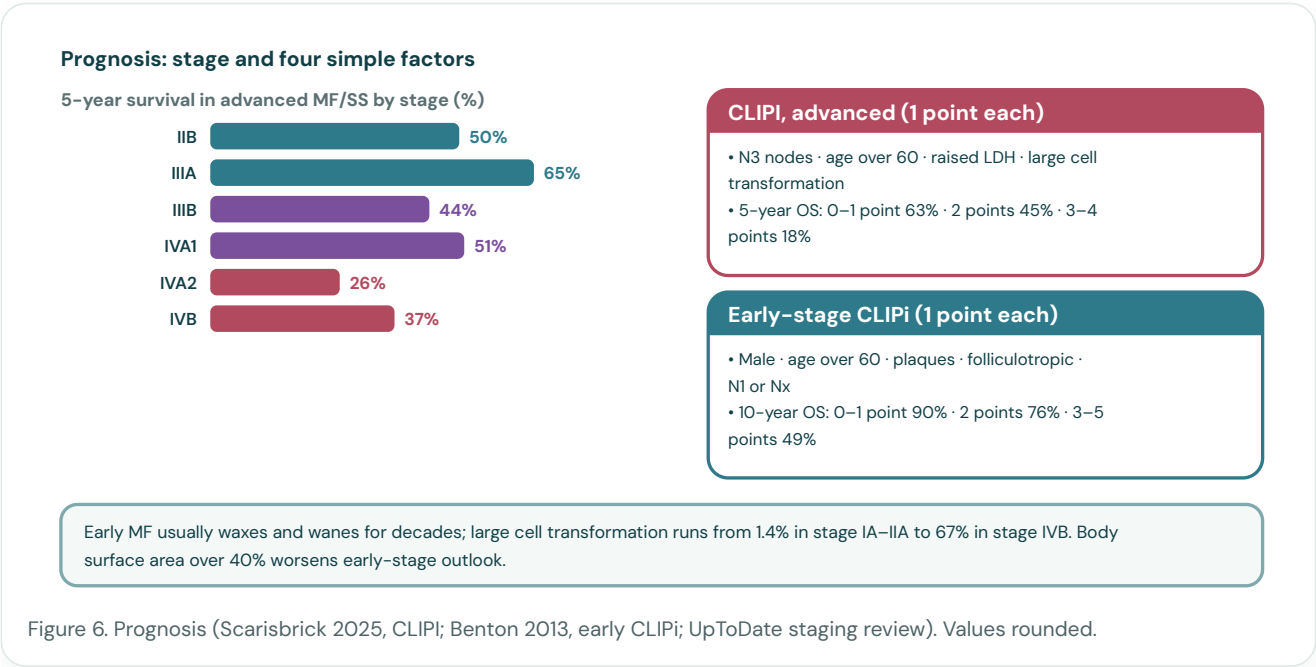
Plain-language myth pages written for patients and families. Send them from the clinic to reinforce what you discussed.

[FNAC alone can reliably diagnose every lymphoma?](#)

[Stage IV lymphoma means the disease is incurable?](#)

## 04 Biology and biomarkers that change treatment

| Marker or feature                                | What it tells you                                                   | What it changes                                                                                |
|--------------------------------------------------|---------------------------------------------------------------------|------------------------------------------------------------------------------------------------|
| CD30 on tumour cells                             | C-ALCL, LyP, many tumour-stage and transformed MF (40–50% of LCT)   | Brentuximab vedotin; even low expression may respond ( <a href="#">ALCANZA<sup>[5]</sup></a> ) |
| Blood class (BO/B1/B2) by flow cytometry + clone | B1 in early MF and B2 in SS carry worse prognosis                   | Systemic treatment needed; ECP or mogamulizumab for blood                                      |
| Large cell transformation (>25% large cells)     | 1.4% in IA–IIA to 67% in IVB; worse survival                        | Treat as advanced (BV); consider allo-HCT                                                      |
| Folliculotropism                                 | Infiltrate around and in hair follicles; deeper than NBUVB reaches  | PUVA or RT for early FMF; tumour-stage FMF as advanced MF                                      |
| TCRγδ, CD8 with epidermotropism and necrosis     | γδ TCL; CD8+ AECTCL                                                 | Aggressive: systemic treatment, transplant referral                                            |
| HAVCR2 (TIM-3) germline variants; HLH            | SPTCL with HLH (17%): 5-year survival 46% vs 91%                    | Urgent HLH work-up and treatment                                                               |
| BCL2, MUM1, FOXP1, IgM, MYD88                    | Leg-type DLBCL rather than PCFCL                                    | Treat as leg-type DLBCL (aggressive B-cell module)                                             |
| Class-switched (IgG) vs IgM PCMZL                | IgM cases resemble MALT lymphoma and spread outside skin more often | Stage IgM cases carefully                                                                      |



### Prognostic scores

- **CLIPi for advanced MF/SS:** N3, age over 60, raised LDH and LCT, 1 point each; 5-year OS 63%, 45% and 18% for 0–1, 2 and 3–4 points ([Scarlsbrick 2025<sup>\[6\]</sup>](#)).
- **Early-stage CLIPi:** male, over 60, plaques, folliculotropic, N1/Nx; 10-year OS 90%, 76% and 49% ([Benton 2013<sup>\[7\]</sup>](#)).

## 05 Local therapy: skin-directed treatment, radiotherapy & surgery

### Skin-directed treatment for mycosis fungoides

- **Topical corticosteroids** (high or super-high potency): first choice for stage IA; response in 94% with 63% complete in T1 disease ([Zackheim 1998](#)<sup>[8]</sup>); expect a response within 3 months; watch atrophy and striae.
- **Topical mechlorethamine**: response in 70–80% of T1 and benefit in 50–70% of T2; the gel was non-inferior to the ointment (CAILS response 59% vs 48%; [Lessin 2013](#)<sup>[9]</sup>). **Topical bexarotene, imiquimod**: for localised, refractory lesions only (topical retinoids are too irritating for large areas).
- **Phototherapy**: NB-UVB or PUVA for IB–IIA patches and thin plaques; any response 88% vs 91%, complete response 62% vs 74% ([Phan 2019](#)<sup>[10]</sup>). Taper slowly after clearing; most relapse when maintenance stops but respond again. PUVA reaches folliculotropic disease better; avoid it after multiple skin cancers.

#### Early-stage MF (IA–IIA): skin-directed treatments and what they achieve

Reported response rates (%), different studies and endpoints



Stage IA: topical steroid first; then mechlorethamine, bexarotene gel, imiquimod or local RT. IB–IIA thin plaques: steroid, mechlorethamine, NB-UVB or PUVA. Thick, widespread, symptomatic plaques: TSEBT (low dose 10–12 Gy, repeatable). Folliculotropic MF needs deeper treatment (PUVA, RT); NB-UVB alone is usually not enough.

**Early aggressive chemotherapy plus electron beam did not improve survival over sequential topical treatment (Kaye 1989). Rotate skin-directed options before systemic drugs.**

Figure 7. Skin-directed therapy (Zackheim 1998; Lessin 2013; Phan 2019; Thomas 2013; Hoppe 2015; Kaye 1989; UpToDate early-stage MF review). Mechlorethamine bar = midpoint of 70–80%.

### Radiotherapy

- **Local RT for MF plaques and tumours**: 8–12 Gy in one to six fractions; a single fraction (mostly 8 Gy) cleared 94% of 270 lesions ([Thomas 2013](#)<sup>[11]</sup>); 4 Gy gives less durable control. **Unilesional MF** (curative): 24–30 Gy. **Symptomatic nodes**: 12–24 Gy, fractionated.
- **TSEBT**: low dose 10–12 Gy over 2–3 weeks (response 88%, complete 27% in pooled phase 2 trials; repeatable; [Hoppe 2015](#)<sup>[12]</sup>, [Kamstrup 2015](#)<sup>[13]</sup>) or conventional 30–36 Gy over 9–10 weeks. For thick, widespread, symptomatic plaques, tumours and erythroderma; follow with topical, light or a retinoid for at least 6 months. Erythema, desquamation and hair loss are common; needs an experienced centre.
- **Indolent cutaneous B-cell lymphoma**: RT for lesions within one field. PCFCL: response-adapted very low dose (2 × 2 Gy, margin 1–1.5 cm), escalating to 24 Gy if needed; in 535 patients (ILROG) complete response was 91% at any dose, but 5-year local relapse-free survival was 96% with >4 Gy vs 73% with ≤4 Gy ([Oertel 2026](#)<sup>[14]</sup>). PCMZL: 24 Gy typical, with a margin ([ILROG 2015](#)<sup>[15]</sup>).

### Surgery

- **Diagnosis:** excisional biopsies for nodules, fat and B-cell lesions.
- **Treatment:** excision for solitary PCFCL or PCMZL in sites unsuitable for RT, unilesional MF, acral CD8+ and CD4+ small/medium T-LPD. Skin relapse after excision is common (about 40% in PCFCL and PCMZL series).

## o6 Systemic therapy: indications & options

### Mycosis fungoides, early stage (IA–IIA)

- **Skin-directed treatment, not systemic:** early aggressive chemotherapy with electron beam gave no survival benefit over sequential topical treatment ([Kaye 1989<sup>\[16\]</sup>](#)).
- **IA:** topical steroid → mechlorethamine, bexarotene gel, imiquimod or local RT; most patients have several skin-directed courses over a lifetime. **IB–IIA thin plaques:** steroid, mechlorethamine, NBUBV or PUVA. **Thick, widespread, symptomatic plaques:** TSEBT.
- **Add a systemic agent** (oral bexarotene or low-dose methotrexate first; interferon, vorinostat, mogamulizumab or BV less often) for extensive or plaque-predominant disease, inadequate control, no access to phototherapy or TSEBT, folliculotropic MF, LCT or B1 blood.
- **Folliculotropic MF:** early form → PUVA, superpotent steroid or local RT (NBUBV alone usually inadequate); tumour form → as stage IIB ([Hodak 2016<sup>\[17\]</sup>](#), [van Santen 2017<sup>\[18\]</sup>](#)). **B1 blood:** treat as stage III with B1.
- **Itch and dry skin:** emollients straight after bathing, topical steroids, antihistamines; gabapentin, mirtazapine or aprepitant for refractory itch.

### Mycosis fungoides, advanced stage (IIB–IV)

#### Advanced MF (IIB–IV): choose by compartment

| Tumours (IIB)                                                                                                                                                                                   | Erythroderma / blood                                                                                                                          | Nodes N3 / viscera                                                                                                                                                      | Aggressive / LCT                                                                                                                                                       |
|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <ul style="list-style-type: none"> <li>• Few tumours: local RT 8–12 Gy + topicals</li> <li>• Generalised: brentuximab vedotin</li> <li>• TSEBT; gemcitabine or liposomal doxorubicin</li> </ul> | <ul style="list-style-type: none"> <li>• Stage III and Sézary: treat like SS</li> <li>• ECP, mogamulizumab, interferon, bexarotene</li> </ul> | <ul style="list-style-type: none"> <li>• BV, romidepsin or pralatrexate</li> <li>• Nodal RT 12–24 Gy if symptomatic</li> <li>• B2 blood: avoid relying on BV</li> </ul> | <ul style="list-style-type: none"> <li>• Repeat biopsy when pace changes</li> <li>• LCT: brentuximab vedotin</li> <li>• Fit, ≤65: allo-HCT at best response</li> </ul> |

#### Randomised evidence (%)

|                                          |     |
|------------------------------------------|-----|
| ALCANZA MF · BV · response ≥4 months     | 50% |
| ALCANZA MF · methotrexate or bexarotene  | 10% |
| MAVORIC · mogamulizumab · blood response | 68% |
| MAVORIC · vorinostat · blood response    | 19% |

ALCANZA (MF, CD30+): median PFS 16.1 vs 3.5 months; tumour-stage response 63% vs 5%; neuropathy common. MAVORIC: median PFS 8 vs 3 months; no visceral responses. Autologous transplant is not used in MF: responses are short. Allogeneic HCT is the only curative option (CUTALLO: median PFS 9 vs 3 months vs no transplant).

Figure 8. Advanced MF (Prince 2017 and Horwitz 2021, ALCANZA; Kim 2018, MAVORIC; de Masson 2023, CUTALLO; UpToDate advanced MF review).

- **IIB, few tumours** (patches and plaques under 10% BSA): local RT plus topical treatment; a complementary systemic agent (methotrexate, bexarotene, interferon) may help.
- **IIB, generalised tumours (≥10% BSA), and LCT:** brentuximab vedotin. In ALCANZA (CD30+ MF and C-ALCL), BV vs methotrexate or bexarotene: MF response lasting ≥4 months 50% vs 10%, tumour-stage 63% vs 5%, median PFS 16.1 vs 3.5 months; 9% grade ≥3 neuropathy; no survival difference ([ALCANZA<sup>\[5\]</sup>](#), [final data<sup>\[19\]</sup>](#)). Without BV: romidepsin, pralatrexate, TSEBT, gemcitabine or liposomal doxorubicin.

- **Stage III (erythroderma, B0–B1) and stage IVA1:** manage as Sézary syndrome (below). **IVA2 MF:** by skin type (erythroderma → as SS; tumours → as IIB/IVB) with RT to symptomatic nodes.
- **IVB (viscera):** BV, romidepsin or pralatrexate (romidepsin 34% overall, 57% blood response in one CTCL series), with RT 8–12 Gy to skin or 12–24 Gy to nodes as needed; gemcitabine (71%; [Zinzani 2000](#)<sup>[20]</sup>) or liposomal doxorubicin (41%; [EORTC 21012](#)<sup>[21]</sup>) also work. Avoid relying on BV for B2 blood.
- **Allogeneic HCT,** the only cure: fit patients up to about 65 with LCT, progressive or refractory disease, done at best response. CUTALLO: median PFS 9 vs 3 months vs no transplant ([CUTALLO](#)<sup>[22]</sup>); CIBMTR 5-year OS 32% ([Lechowicz 2014](#)<sup>[23]</sup>); EBMT 7-year OS 44%, with most survivors in CR ([Duarte 2014](#)<sup>[24]</sup>). No autologous HCT (short responses). BV is a safe bridge; mogamulizumab with caution before transplant.

## Sézary syndrome

### Sézary syndrome: three criteria, then treatment by visceral disease

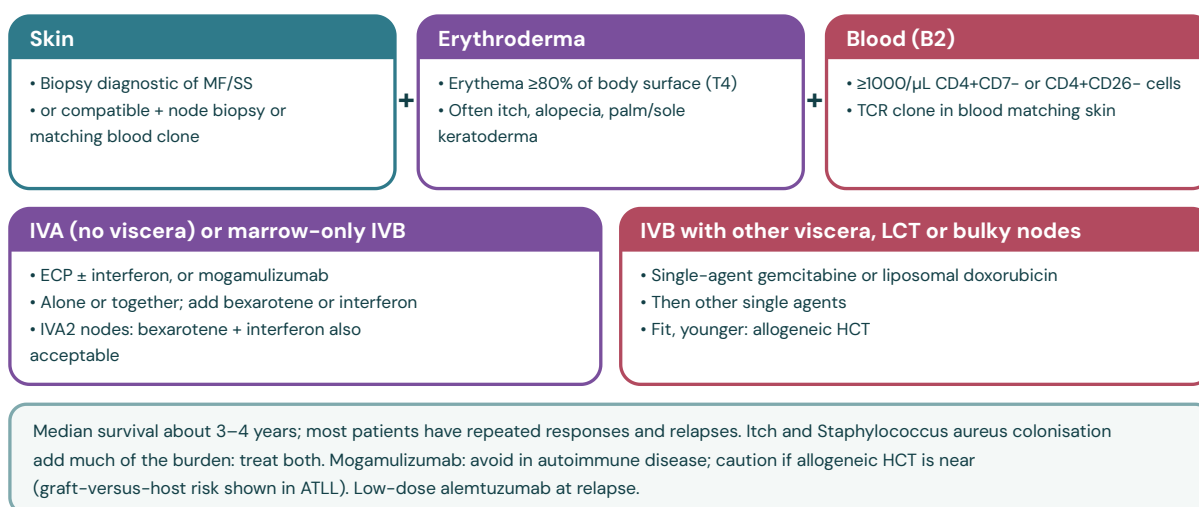


Figure 9. Sézary syndrome (Olsen 2022; Kim 2018, MAVORIC; UpToDate SS diagnosis and treatment reviews).

- **Stage IVA (no viscera), and marrow-only IVB:** extracorporeal photopheresis (with interferon) or mogamulizumab, alone, together or with bexarotene or interferon. ECP is slow and needs a specialised centre; mogamulizumab is avoided in autoimmune disease and is weaker for bulky nodes, tumours and LCT. IVA2: bexarotene plus interferon is also acceptable.
- **MAVORIC** (previously treated MF and SS): mogamulizumab vs vorinostat, median PFS 8 vs 3 months; responses in blood 68% vs 19%, skin 42% vs 16%, nodes 17% vs 4%; SS response 37% vs 2% ([MAVORIC](#)<sup>[25]</sup>).
- **IVB with other viscera, LCT, rapid progression or bulky nodes:** single-agent gemcitabine or liposomal doxorubicin rather than combination chemotherapy; then the other agent; allogeneic HCT for younger fit patients. Relapse: sequential single agents, low-dose alemtuzumab, pembrolizumab, trials.
- **Supportive care:** itch (emollients, steroids, doxepin at night, gabapentin or pregabalin, mirtazapine, aprepitant or naltrexone); culture crusted erosions; dilute bleach or chlorhexidine baths with topical antibiotic; systemic antibiotics (think MRSA) for infection; watch for disseminated herpes in erythroderma.

## CD30+ lymphoproliferative disorders: lymphomatoid papulosis and primary cutaneous ALCL

## Primary cutaneous CD30+ lymphoproliferative disorders: one spectrum

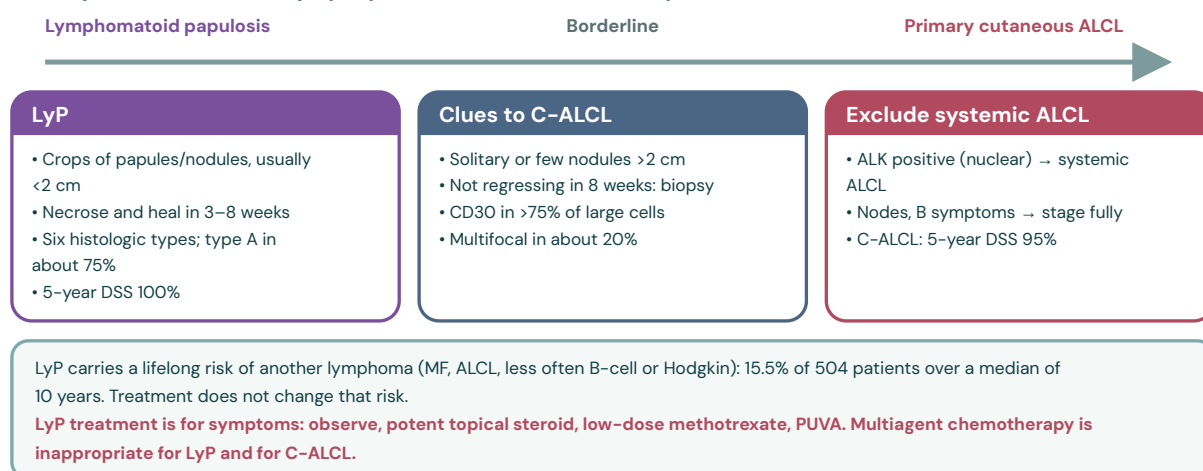
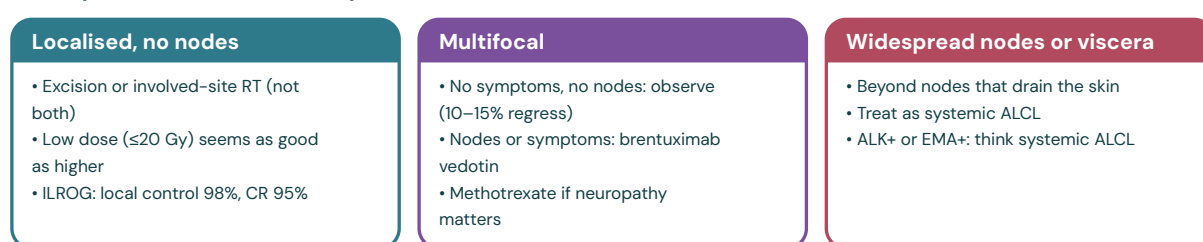


Figure 10. CD30+ disorders (Kempf 2011 consensus; Melchers 2020; WHO–EORTC 2018; UpToDate LyP review).

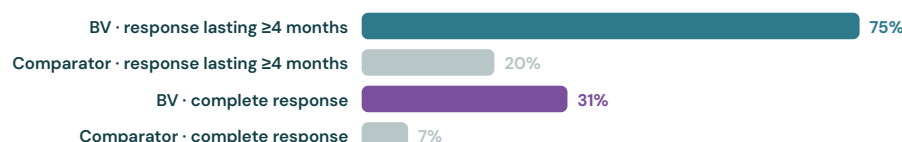
- **LyP:** treatment relieves symptoms but does not change the course or the risk of a second lymphoma. Few lesions → observe or a super-potent topical steroid. Many, symptomatic or on face and hands → low-dose weekly methotrexate (with folic acid; often needs maintenance; liver and blood monitoring), then PUVA (or NBUBV/UVA1) if methotrexate fails or is contraindicated. Multiagent chemotherapy is inappropriate ([Kempf 2011](#)<sup>[26]</sup>).
- **Children with LyP:** often no treatment; topical steroids or NBUBV when needed; low-dose methotrexate second.
- **Biopsy any lesion >2 cm that has not regressed in 8 weeks** to look for primary cutaneous ALCL; nuclear ALK or nodes → systemic ALCL (T-cell module).
- **Lifelong follow-up:** 15.5% of 504 LyP patients developed another haematological cancer (mostly MF or ALCL) over a median of 10 years; deaths from it are uncommon ([Melchers 2020](#)<sup>[27]</sup>, [Bekkenk 2000](#)<sup>[28]</sup>).

## Primary cutaneous anaplastic large cell lymphoma

## Primary cutaneous ALCL: treat by extent, and do not overtreat



## ALCANZA, 31 patients with C-ALCL: BV vs methotrexate or bexarotene (%)



Median PFS 28 vs 5 months. CHOP-type chemotherapy gives high but short responses (CR 92%, 62% relapsed at a median of 4 months) with real toxicity. Outlook: disease-related 5-year survival 95% (Dutch registry); skin relapse in about 40%; leg lesions 5-year DSS 76% vs 96% elsewhere; ≥5 lesions at onset: more spread and deaths.

Figure 11. Primary cutaneous ALCL (Million 2016, ILROG; Prince 2017 and Horwitz 2021, ALCANZA; Bekkenk 2000; Hapgood 2017; Melchers 2018; UpToDate PC-ALCL review).

- **Who and how:** about 8–10% of cutaneous lymphomas; median age 61, slightly more men (SEER). Solitary or grouped nodules, usually on the upper body, growing over weeks to months and often ulcerating; up to half regress partly, rarely completely. About a quarter had two or more sites in the Dutch registry.
- **Pathology:** dense dermal sheets of large anaplastic cells with CD30 on  $\geq 75\%$ ; most CD4+ with variable loss of CD2, CD3, CD5; ALK negative and usually EMA negative (both often positive in systemic ALCL); DUSP22::IRF4 rearrangement in up to half, with an indolent course. Exclude ATLL (CD30+ anaplastic, ALK-: test HTLV-1), CD30+ transformed MF, Hodgkin (CD15+) and reactive CD30+ infiltrates.
- **Staging:** examination and blood tests; CT or PET-CT if extracutaneous disease is suspected; biopsy enlarged draining nodes (inflammation is common); marrow only for multifocal tumours, cytopenias or proven extracutaneous disease. Involvement of draining nodes alone does not worsen prognosis.
- **Localised, no node involvement:** excision or involved-site RT, not both, and no chemotherapy; low-dose RT ( $\leq 20$  Gy) appears as effective as higher doses. ILROG series (56 patients): local control 98%, complete response 95%, responses from 6 Gy ([Million 2016](#)<sup>[29]</sup>, [Smith 2017](#)<sup>[30]</sup>). **With involved draining nodes:** ISRT to skin and nodes.
- **Multifocal:** asymptomatic, no nodes → observe (10–15% regress; wait 4–6 weeks for a large lesion, then excise or irradiate if it persists); nodes or systemic symptoms → brentuximab vedotin. In ALCANZA (31 C-ALCL patients) BV beat methotrexate or bexarotene: response  $\geq 4$  months 75% vs 20%, CR 31% vs 7%, median PFS 28 vs 5 months; neuropathy improved or resolved in 86% with longer follow-up ([ALCANZA](#)<sup>[5]</sup>, [final data](#)<sup>[19]</sup>). Oral methotrexate is an alternative; CHOP-type chemotherapy gives brief responses ([Melchers 2018](#)<sup>[31]</sup>).
- **Widespread nodal or visceral disease** (beyond nodes draining the skin): treat as systemic ALCL (T-cell module).
- **Relapse:** a localised skin relapse → observe first (some regress); if it persists beyond about 2 months or causes symptoms, RT, excision, or super-potent topical or intralesional steroid; multiple skin relapses → BV again if not given in the last 6 months, no progression on it and no neuropathy (88% response on retreatment in systemic ALCL), or methotrexate or bexarotene; widespread or after BV → sequential single agents.
- **Outlook:** disease-related 5-year survival 95% (Dutch registry; [Bekkenk 2000](#)<sup>[28]</sup>), yet skin relapse in about 40% within 5 years and systemic lymphoma in 14% ([Hapgood 2017](#)<sup>[32]</sup>); SEER 5- and 10-year OS 81% and 62% ([SEER](#)<sup>[33]</sup>). Worse with leg lesions (5-year DSS 76% vs 96%; [Miyagaki 2023](#)<sup>[34]</sup>), five or more lesions (extracutaneous relapse 56% vs 20%), age over 60 and chemotherapy use.

## SPTCL and the rare cutaneous T-cell lymphomas

**Rare cutaneous T-cell lymphomas: two indolent, two aggressive, one in the fat**

|                                                                                                                  |                                                                                                     |                                                                                                                           |
|------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------------------------------------------|
| <b>SPTCL</b><br>Fat nodules, legs and arms, young women; autoimmune disease in up to 20%; 5-year OS about 80%    | <b>SPTCL with HLH</b><br>HLH in 17%; 5-year survival 46% vs 91% without; germline HAVCR2 (TIM-3)    | <b>γδ T-cell lymphoma</b><br>Ulcerating plaques and tumours on limbs; chemo-resistant; median survival about 15 months    |
| <b>CD8+ AECTCL</b><br>Eruptive necrotic papules and tumours; median survival under 2 years; allo-HCT in selected | <b>Acral CD8+ T-LPD</b><br>Single nodule, usually the ear; excision or RT; 10% regress after biopsy | <b>CD4+ small/medium T-LPD</b><br>Solitary face/neck/upper trunk nodule; 8–12% of CTCL; intralesional steroid or excision |

SPTCL needs a deep (excisional) biopsy including fat; shave biopsies miss it. Screen every febrile, cytopenic patient for HLH (ferritin, triglycerides, fibrinogen).

**Large, fast-growing generalised lesions with >30% large cells are not CD4+ small/medium T-LPD: classify as PTCL, NOS. SPTCL treatment is not yet covered in this module.**

Figure 12. Rare subtypes (Willemze 2008; Gayden 2018; Toro 2003; Berti 1999; Kempf 2022; UpToDate SPTCL and rare-subtype reviews).

- **SPTCL**: mostly indolent (5-year OS about 80%); spread outside the fat is rare and some cases regress. Prognosis is driven by HLH (5-year survival 46% vs 91%), linked to germline HAVCR2 (TIM-3) variants ([Willemze 2008](#)<sup>[35]</sup>, [Gayden 2018](#)<sup>[36]</sup>). IPI and PIT put most patients in low risk and miss the HLH effect.
- **Primary cutaneous γδ T-cell lymphoma**: ulcerated plaques and tumours on the limbs, often with fat involvement (worse); resistant to multiagent chemotherapy; median survival about 15 months ([Toro 2003](#)<sup>[37]</sup>).
- **CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma**: eruptive necrotic papules and tumours; chemotherapy responses disappointing; median survival under 2 years; durable remissions after allogeneic HCT in selected patients ([Berti 1999](#)<sup>[38]</sup>). Distinguish from CD8+ MF, which behaves like ordinary MF.
- **SPTCL mimics**: lupus panniculitis (lupus profundus) favours the face, scalp, upper arms, breasts, upper trunk, buttocks and thighs, is usually painful, heals with marked atrophy, may have discoid lupus on top, shows lymphoid follicles and has no clonal TCR; "cytotoxic histiocytic panniculitis" (macrophages eating blood cells, "beanbag cells") can occur with SPTCL or without any lymphoma.
- **Acral CD8+ T-LPD** (usually the ear) and **CD4+ small/medium T-LPD** (solitary face, neck or upper trunk nodule; 8–12% of CTCL): excision, RT or intralesional steroid; some regress after biopsy ([Kempf 2022](#)<sup>[39]</sup>).

**SPTCL treatment: not yet covered in this module**

- Treatment of SPTCL will be added after review of a dedicated source. Plan treatment with the lymphoma team; manage suspected HLH urgently (section O6 and Table D).

## Primary cutaneous follicle centre and marginal zone lymphomas

**Primary cutaneous B-cell lymphomas: two indolent, one aggressive****PCFCL (most common)**

- Firm nodules on scalp, head, trunk
- BCL6+, BCL2-; not graded
- 5-year OS about 95%; leg site worse

**PCMZL**

- Red-violet papules/nodules, trunk and arms
- Multifocal often; BCL2+, BCL6-
- 5-year survival 98–100%

**PCDLBCL, leg type**

- Older women, lower legs
- BCL2, MUM1, FOXP1, IgM; MYD88
- Aggressive: see aggressive module

RT for limited indolent PCBCL (ILROG, 535 patients): 5-year local relapse-free survival (%)



Same complete response (91%) at both doses, more local relapse after ≤4 Gy. Localised: RT (PCFCL: response-adapted 4 Gy, up to 24 Gy; PCMZL: 24 Gy typical). Extensive symptomatic PCFCL: single-agent rituximab. PCMZL multifocal and quiet: observe; symptomatic: intralesional steroid, low-dose RT or excision. Chemotherapy is rarely needed.

Figure 13. Cutaneous B-cell lymphomas (Senff 2008; Oertel 2026, ILROG; UpToDate PCFCL and PCMZL reviews).

- **PCFCL** (EORTC/ISCL, ILROG, NCCN; [Senff 2008](#)<sup>[40]</sup>): localised and symptomatic or cosmetically troublesome → RT; not troublesome → topical or intralesional steroid or observation. Lesions beyond one field → RT to up to three symptomatic sites, steroids for the rest. Extensive symptomatic disease → single-agent rituximab; combination chemotherapy only for very extensive disease or rituximab failure. About 10% spread outside the skin over 10 years.
- **PCMZL**: lesions in one field → RT (24 Gy) or excision; asymptomatic multifocal → observe; symptomatic multifocal → intralesional triamcinolone, low-dose RT or excision rather than chemotherapy. Five-year survival 98–100%. Borrelia testing where relevant (Europe).
- **Relapse (both)**: about a third relapse after RT, mostly in skin. Outside the field → RT; inside the field → excision or rituximab. Extracutaneous PCFCL → treat as follicular lymphoma (indolent B-cell module).

**India now (October 2026)**

- Indian data are web-verified: a South Indian series (MF 57%, ATLL 17% of cutaneous T-cell lymphomas; Khader 2016) and the KEM Mumbai hypopigmented MF series (mimicking Hansen's disease).
- Availability and cost in India of mechlorethamine gel, topical and oral bexarotene, PUVA/NBUVB units, TSEBT, extracorporeal photopheresis, mogamulizumab, brentuximab vedotin, romidepsin and pralatrexate, and access to allogeneic transplant, to be confirmed by the treating team.

**SHARE WITH PATIENTS: TREATMENT AND RELAPSE**

Plain-language myth pages written for patients and families. Send them from the clinic to reinforce what you discussed.

Chemotherapy is needed immediately for every lymphoma?

Radiotherapy has no role in lymphoma?

Immunotherapy and targeted therapy are the same as chemotherapy?

If lymphoma returns after chemotherapy, no effective treatment remains?

**Quick reference (also as a printable PDF on the Printable Teaching Resources page)**

**Table A. Diagnosis and staging tests and what they change**

| Test                                                                                                                                     | When                                                                          | What it changes                                   | Pitfalls                                                              |
|------------------------------------------------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------|---------------------------------------------------|-----------------------------------------------------------------------|
| Skin punch biopsy ≥4 mm (most indurated lesion); often several over time                                                                 | Every suspected MF                                                            | ISCL/EORTC score; variant (folliculotropic, LCT)  | Topicals in the last 2–4 weeks blur the picture                       |
| Excisional or deep incisional biopsy including fat                                                                                       | Nodules, SPTCL, B-cell lymphomas, LyP (2 fresh lesions)                       | Entity                                            | Shave biopsies are inadequate                                         |
| IHC: CD2, CD3, CD4, CD5, CD7, CD8, CD30, ALK, TCRβF1/γδ, CD56, cytotoxic markers; B-cell panel with BCL2, BCL6, MUM1, CD10, light chains | Every biopsy                                                                  | Names the entity; CD30 → BV option                | Read with the clinical picture: pseudolymphomas mimic all of these    |
| TCR (or IgH) clonality, skin and blood                                                                                                   | Equivocal histology; erythroderma; staging B and N                            | 1 ISCL point; B2 needs a skin-matched clone       | Clones occur in benign disease and with age                           |
| Blood flow cytometry (CD4+CD7-, CD4+CD26-, TRBC1)                                                                                        | Every MF/SS staging                                                           | BO/B1/B2; Sézary syndrome                         | Lymphopenia: use percentages                                          |
| CBC, chemistry, LDH; HTLV-1 serology if from an endemic area or flower cells/high calcium                                                | Baseline                                                                      | CLIP1; ATLL excluded                              | ATLL was 17% of cutaneous T-cell lymphomas in one South Indian series |
| CT or PET-CT                                                                                                                             | MF with T2b, T3, T4, nodes >1.5 cm, LCT or B2; all non-MF cutaneous lymphomas | N and M stage; primary vs secondary skin lymphoma | Chest X-ray if no CT/PET in limited MF                                |
| Node excision; marrow biopsy only if cytopenias or equivocal PET                                                                         | Enlarged node; suspected systemic disease                                     | N3 (IVA2); M1                                     | FNA cannot grade nodal involvement                                    |

**Table B. Treatment by setting**

| Setting                                 | Standard choice                                                                   | Key evidence                                                                               | Notes                                              |
|-----------------------------------------|-----------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------|----------------------------------------------------|
| MF IA                                   | Potent topical steroid; then mechlorethamine, bexarotene gel, imiquimod, local RT | <a href="#">Zackheim 1998</a> <sup>[8]</sup>                                               | Rotate skin-directed options before systemic drugs |
| MF IB–IIA, thin plaques                 | Steroid, mechlorethamine, NBUBV or PUVA                                           | <a href="#">Phan 2019</a> <sup>[10]</sup>                                                  | PUVA: more CR; both need maintenance tapering      |
| MF IB–IIA, thick widespread plaques     | TSEBT (low dose 10–12 Gy), then adjuvant topical, light or retinoid               | <a href="#">Hoppe 2015</a> <sup>[12]</sup>                                                 | Add systemic agent if B1, folliculotropic or LCT   |
| MF IIB, few tumours                     | Local RT 8–12 Gy + topicals                                                       | <a href="#">Thomas 2013</a> <sup>[11]</sup>                                                | Electrons or low-energy x-rays                     |
| MF IIB, generalised tumours; LCT        | Brentuximab vedotin                                                               | <a href="#">ALCANZA</a> <sup>[5]</sup>                                                     | Neuropathy; TSEBT after response                   |
| MF III and Sézary IVA                   | ECP ± interferon, or mogamulizumab                                                | <a href="#">MAVORIC</a> <sup>[25]</sup>                                                    | Treat itch and Staphylococcus too                  |
| MF IVB; SS with M1b, LCT or bulky nodes | BV, romidepsin or pralatrexate (MF); gemcitabine or liposomal doxorubicin (SS)    | <a href="#">Zinzani 2000</a> <sup>[20]</sup> , <a href="#">EORTC 21012</a> <sup>[21]</sup> | RT 12–24 Gy to symptomatic nodes                   |
| Advanced MF/SS, fit, ≤65                | Allogeneic HCT at best response                                                   | <a href="#">CUTALLO</a> <sup>[22]</sup>                                                    | No autologous HCT                                  |
| LyP                                     | Observe or topical steroid; low-dose methotrexate; PUVA                           | <a href="#">Kempf 2011</a> <sup>[26]</sup>                                                 | No multiagent chemotherapy                         |

| Setting                                   | Standard choice                                                      | Key evidence                                 | Notes                                       |
|-------------------------------------------|----------------------------------------------------------------------|----------------------------------------------|---------------------------------------------|
| PCFCL or PCMZL, localised                 | RT (PCFCL 4 Gy response–adapted to 24 Gy; PCMZL 24 Gy) or excision   | <a href="#">ILROG 2026</a> <sup>[14]</sup>   | Leg site PCFCL: worse outlook               |
| PCFCL extensive; PCMZL multifocal         | PCFCL: rituximab; PCMZL: observe, intralesional steroid, low-dose RT | <a href="#">Senff 2008</a> <sup>[40]</sup>   | Chemotherapy rarely                         |
| Acral CD8+ or CD4+ small/medium T-LPD     | Excision, RT or intralesional steroid                                | <a href="#">Kempf 2022</a> <sup>[39]</sup>   | Behave like pseudolymphoma                  |
| Primary cutaneous ALCL, localised         | Excision or involved-site RT (≤20 Gy appears enough)                 | <a href="#">Million 2016</a> <sup>[29]</sup> | No added RT after excision; no chemotherapy |
| C-ALCL, multifocal with nodes or symptoms | Brentuximab vedotin (methotrexate if neuropathy)                     | <a href="#">ALCANZA</a> <sup>[5]</sup>       | Asymptomatic multifocal: observe first      |
| SPTCL                                     | Not yet covered in this module                                       | —                                            | Plan with the lymphoma team; HLH is urgent  |

**Table C. Systemic options: evidence and best fit**

| Option                               | Best for                                                               | Evidence                                                                                                         | Watch for                                            |
|--------------------------------------|------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------|------------------------------------------------------|
| Brentuximab vedotin                  | CD30+ MF (tumours, LCT, nodes); multifocal C-ALCL                      | <a href="#">ALCANZA</a> <sup>[5]</sup> : MF response ≥4 months 50% vs 10%; C-ALCL 75% vs 20%, PFS 28 vs 5 months | Peripheral neuropathy; not relied on for B2 blood    |
| Mogamulizumab                        | Blood and erythrodermic skin (SS, stage III)                           | <a href="#">MAVORIC</a> <sup>[25]</sup> : PFS 8 vs 3 months; blood response 68% vs 19%                           | Avoid in autoimmune disease; caution before allo-HCT |
| Extracorporeal photopheresis         | Erythroderma, blood involvement; combine with interferon or bexarotene | Observational series (response in skin common, CR uncommon)                                                      | Specialised centres; slow responses; venous access   |
| Bexarotene, methotrexate, interferon | Patch/plaque and itch in advanced MF; add-on to skin-directed therapy  | Less active than BV in ALCANZA                                                                                   | Bexarotene: triglycerides, thyroid                   |
| Romidepsin, pralatrexate             | Tumours, nodes; MF IVB                                                 | Phase 2 (romidepsin 34% overall, 57% blood response)                                                             | Romidepsin hard for frail patients                   |
| Gemcitabine, liposomal doxorubicin   | Aggressive or visceral MF/SS                                           | <a href="#">Zinzani 2000</a> <sup>[20]</sup> (71%); <a href="#">EORTC 21012</a> <sup>[21]</sup> (41%)            | Myelosuppression                                     |
| Rituximab (single agent)             | Extensive PCFCL; in-field relapse of PCFCL/PCMZL                       | Small series; responses often transient                                                                          | Skin relapse is common                               |
| Allogeneic HCT                       | Advanced MF/SS, fit, best response                                     | <a href="#">CUTALLO</a> <sup>[22]</sup> ; <a href="#">EBMT</a> <sup>[24]</sup> (7-year OS 44%)                   | Non-relapse mortality about 20%                      |

**Table D. Managing problems during treatment**

| Problem       | What to do                                                                                                                                                 | Notes                                         |
|---------------|------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------|
| Itch (MF, SS) | Emollients after bathing, topical steroid, antihistamines; doxepin at night, gabapentin or pregabalin; mirtazapine, aprepitant or naltrexone if refractory | Itch tracks stage; a new infection worsens it |
|               |                                                                                                                                                            |                                               |

| Problem                                         | What to do                                                                                                                                                 | Notes                                                     |
|-------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------|
| Staphylococcus aureus colonisation or infection | Culture crusted or weeping erosions; dilute bleach baths or chlorhexidine plus topical antibiotic; systemic antibiotics (consider MRSA) for real infection | Clearing infection often improves the lymphoma skin signs |
| Herpes simplex or zoster in erythroderma        | Recognise and treat promptly                                                                                                                               | Risk of disseminated infection                            |
| Topical steroid side effects                    | Watch for atrophy and striae; care near the eyes                                                                                                           | Response by 3 months, or change                           |
| Phototherapy and RT late effects                | Skin cancer and cataract surveillance after PUVA; RT dermatitis, alopecia                                                                                  | Avoid PUVA after many skin cancers                        |
| Brentuximab neuropathy                          | Dose delay, reduction or stop (half needed a change in ALCANZA)                                                                                            | 41% still had grade $\leq 2$ neuropathy at long follow-up |
| HLH (SPTCL, $\gamma\delta$ TCL)                 | Ferritin, triglycerides, fibrinogen in a febrile cytopenic patient; urgent haematology                                                                     | Main adverse factor in SPTCL                              |

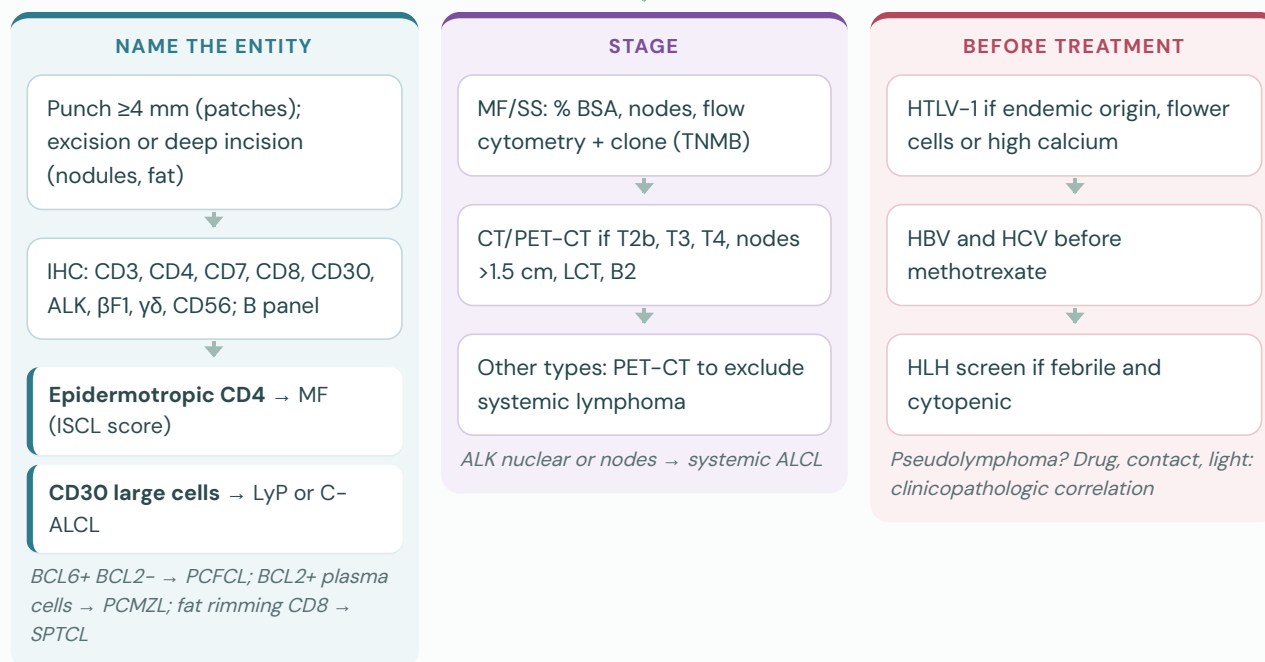
## 07 Management algorithms

Five decision paths pull together sections 02–06.

### ALGORITHM 1

### Suspected cutaneous lymphoma: to an entity and a plan

**Persistent patches in covered skin, pale patches, erythroderma, nodules, self-healing papules or fat nodules** → stop topicals 2–4 weeks → biopsy before steroids or antifungals

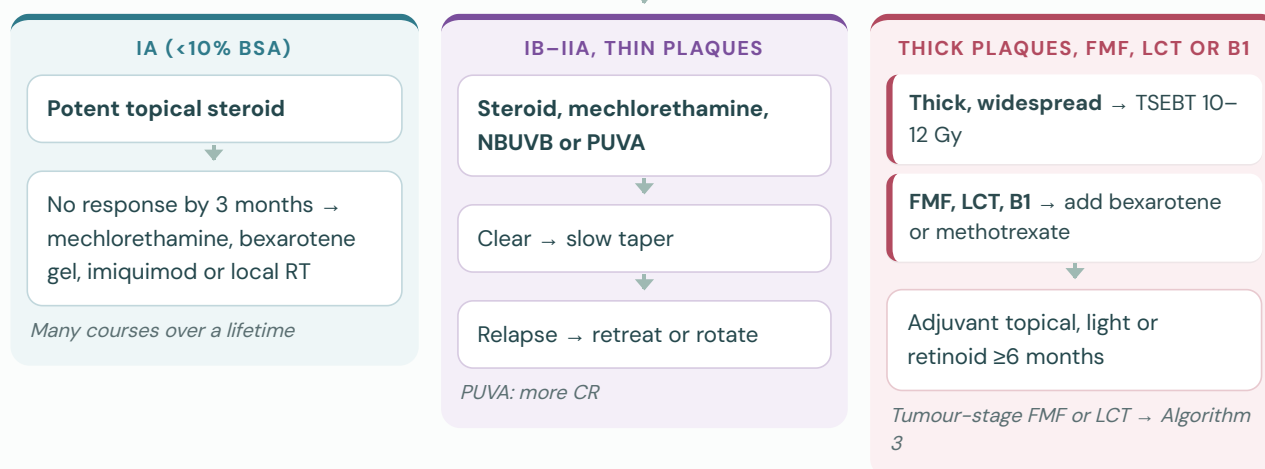


All: clinicopathologic correlation · repeat biopsies when unsure · a clone or CD30 alone is not a diagnosis.

### ALGORITHM 2

### Mycosis fungoides, early stage (IA–IIA)

**MF confirmed, stage IA–IIA** → lesion type, % BSA, folliculotropism, blood class

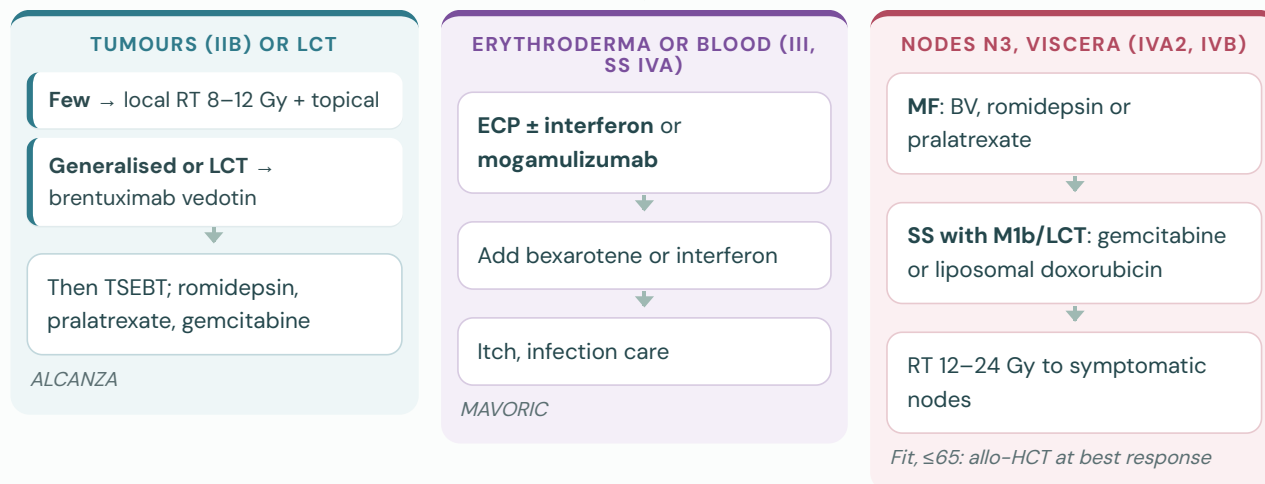


All: emollients and itch control · no early chemotherapy (no survival gain) · biopsy again if the pace changes.

## ALGORITHM 3

## Advanced mycosis fungoides and Sézary syndrome

**Stage IIB–IV** → which compartment dominates: tumours, erythroderma/blood, nodes, viscera?

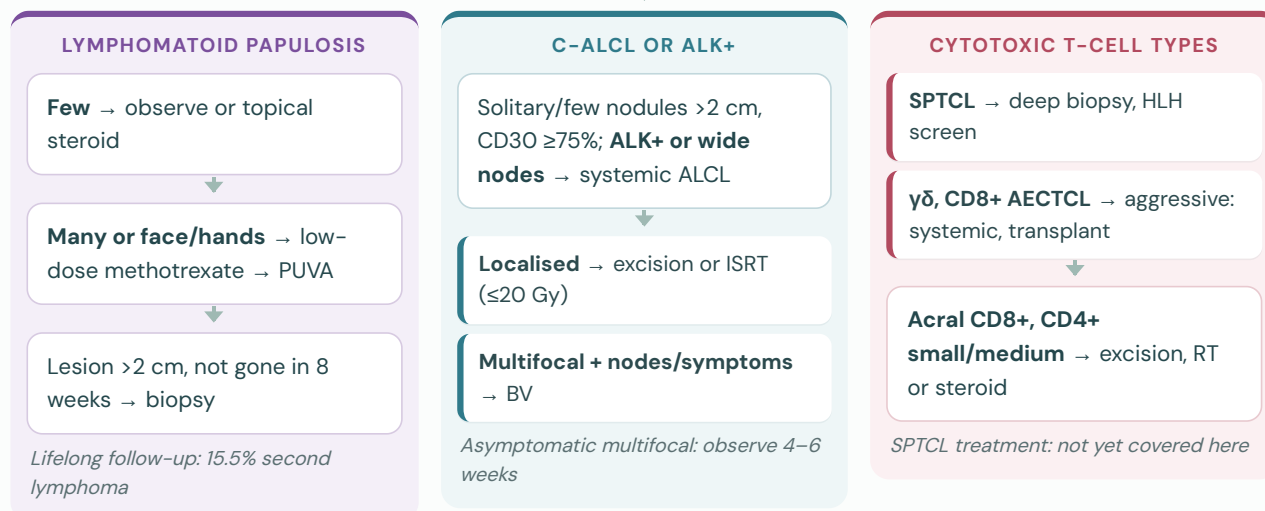


All: multidisciplinary team · maintenance after response · trials · allogeneic HCT is the only cure; no autologous HCT.

## ALGORITHM 4

## CD30+ disorders and rare cutaneous T-cell lymphomas

**CD30+ or cytotoxic infiltrate in the skin** → clinical course, size, ALK, phenotype



All: no multiagent chemotherapy for LyP or C-ALCL · stage fully when ALK+ · HLH is the danger in SPTCL.

## ALGORITHM 5

## Primary cutaneous follicle centre and marginal zone lymphoma

**PCFCL or PCMZL confirmed; PET-CT negative → how many lesions, where, symptomatic?**

## ONE RT FIELD

**Symptomatic** → RT (PCFCL 4 Gy → 24 Gy; PCMZL 24 Gy) or excision

**Not troublesome** → steroid or observe

*ILROG: >4 Gy better local control*

## SEVERAL SITES

**PCFCL:** RT to ≤3 symptomatic sites + steroids

**PCMZL:** observe if quiet; intralesional steroid, low-dose RT or excision

*No chemotherapy*

## EXTENSIVE OR RELAPSE

**Extensive PCFCL** → rituximab

**Relapse outside field** → RT;  
**inside** → excision or rituximab

*Spread beyond skin → nodal lymphoma pathway*

All: about a third relapse in skin · visits every 3 months for 2 years · no routine scans or blood tests.

## o8 Survivorship, follow-up & late effects

### A simple follow-up rhythm for cutaneous lymphomas

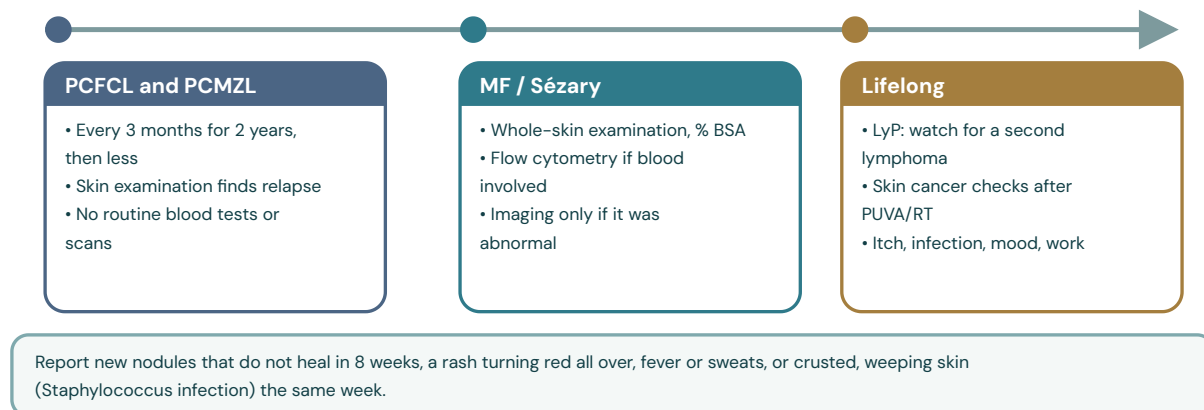


Figure 14. Follow-up rhythm (Terhorst 2012; UpToDate PCFCL, PCMZL, LyP and MF reviews). Individualise.

- **Indolent B-cell lymphomas:** every 3 months for 2 years, then as needed; in one study all 23 relapses of PCFCL were found by examination, none by blood tests or scans ([Terhorst 2012<sup>\[41\]</sup>](#)). **LyP:** lifelong review because of the second-lymphoma risk.
- **MF/SS:** whole-skin examination with % BSA, nodes and organs, CBC, chemistry, LDH, flow cytometry; repeat imaging only if it was abnormal and would change management.
- **Late effects and risks:** skin cancers and cataracts after PUVA and radiation; steroid atrophy; neuropathy after BV; more second lymphomas after MF/SS and LyP; infections (Staphylococcus, herpes).
- **Living with it:** itch affects sleep and mood (itch scores rise with stage); support, counselling and honest discussion of a chronic, relapsing disease.

### Relapse is common, but rarely dangerous, in most cutaneous lymphomas

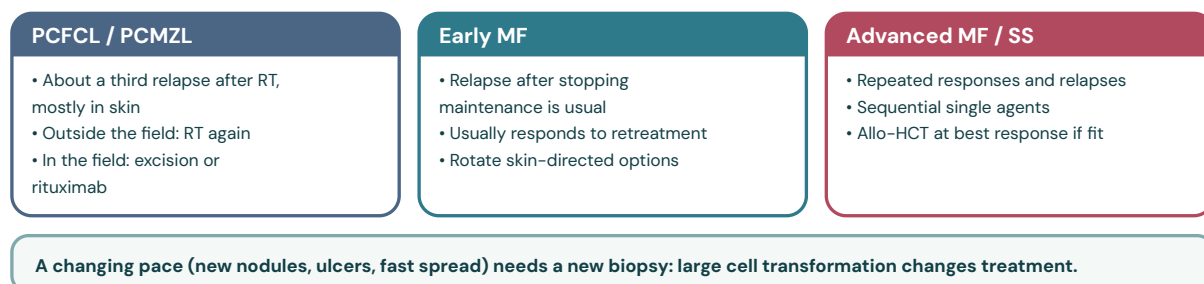


Figure 15. Relapse patterns (UpToDate PCFCL, PCMZL, early and advanced MF and SS reviews).

### SHARE WITH PATIENTS: LIFE AFTER TREATMENT

Plain-language myth pages written for patients and families. Send them from the clinic to reinforce what you discussed.

#### Lymphoma survivors can't return to normal life?

## 09 Landmark trials

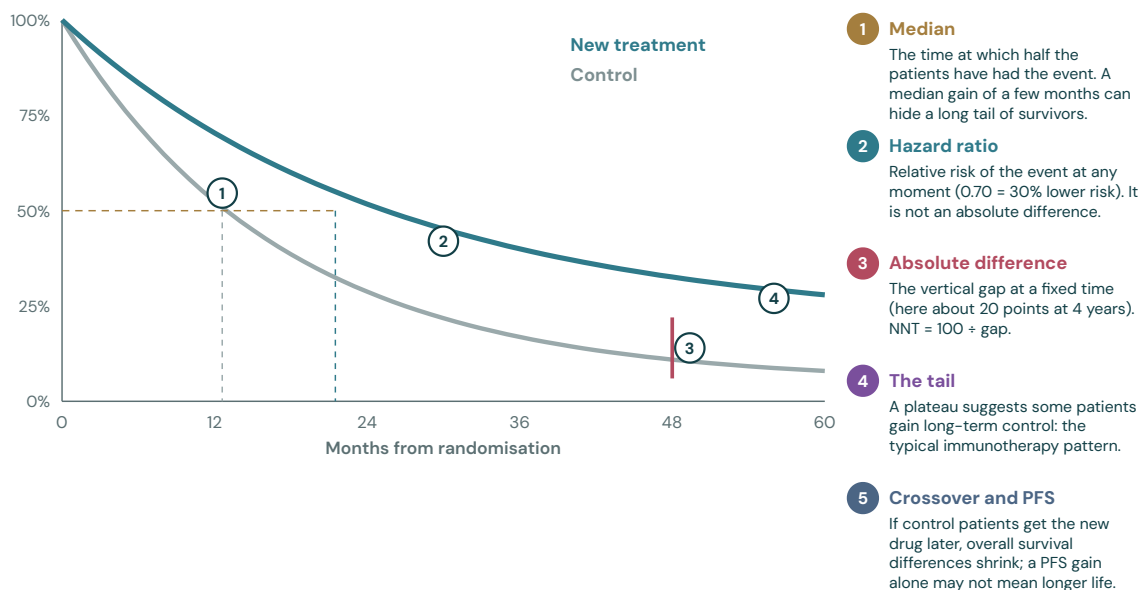


Figure 16. How to read a survival curve (illustrative). Use the absolute gap at a fixed time and overall survival, not the median or PFS alone, when weighing benefit against toxicity and cost.

### The trials behind today's choices

#### ALCANZA · MF and C-ALCL

BV vs methotrexate/bexarotene · response  $\geq 4$  months



#### MAVORIC · MF and SS

Mogamulizumab vs vorinostat · median PFS (months)



#### CUTALLO · advanced CTCL

Allo-HCT vs no transplant · median PFS (months)



#### Phototherapy meta-analysis

PUVA vs NB-UVB · complete response



#### Mechlorethamine gel trial

Gel vs ointment · CAILS response



#### ILROG · indolent PCBCCL

$>4$  Gy vs  $\leq 4$  Gy · 5-year local RFS



Grey = comparator. Endpoints and scales differ (% or months); compare within a trial, not across trials.

Figure 17. Landmark results as reported in the UpToDate MF, SS, early-stage MF and PCFCL reviews.

- [ALCANZA](#)<sup>[5]</sup>: BV vs methotrexate or bexarotene in CD30+ MF and C-ALCL → better and longer responses → BV for tumours and LCT.
- [MAVORIC](#)<sup>[25]</sup>: mogamulizumab vs vorinostat → longer PFS, strong blood responses → mogamulizumab for Sézary and erythrodermic MF.
- [CUTALLO](#)<sup>[22]</sup>: allogeneic HCT vs no transplant in advanced CTCL (by donor availability) → longer PFS.
- [Kaye 1989](#)<sup>[16]</sup>: chemotherapy plus electron beam vs topical therapy first → no survival gain → skin-directed first in early MF.
- [Phan 2019](#)<sup>[10]</sup> and [Lessin 2013](#)<sup>[9]</sup>: the evidence for phototherapy and topical mechlorethamine.

- [ILROG 2026](#)<sup>[14]</sup>: 535 patients; same CR at  $\leq 4$  Gy, better local control above 4 Gy.

## 10 Practice pearls & pitfalls

- Persistent patches on the buttocks, groins and trunk that change size and shape: biopsy, after 2–4 weeks off topicals.
- Pale patches in a child or young adult that do not behave like leprosy, vitiligo or pityriasis alba: think hypopigmented MF.
- Erythroderma in an older adult: flow cytometry (CD4+CD7-/CD26-) and a TCR clone in blood and skin before calling it a drug rash or eczema.
- A T-cell clone or CD30+ cells alone do not make a lymphoma: correlate with the clinical picture.
- Early MF is a chronic skin disease for most people: skin-directed treatment, not chemotherapy.
- New nodules in long-standing plaques: rebiopsy for large cell transformation (BV changes the outlook).
- Self-healing papules with large CD30+ cells: lymphomatoid papulosis; never CHOP.
- ALK-positive CD30+ skin tumour: stage fully for systemic ALCL.
- Fat nodules with fever and cytopenias: SPTCL with HLH until proven otherwise; deep biopsy and ferritin.
- Cutaneous B-cell lymphoma on the scalp or trunk: low-dose RT, excision or steroid; chemotherapy is rarely needed.

## 11 Case vignettes: lines of management

Eight cases from the clinic.

### CASE 1 Hypopigmented MF · teenager

#### Pale patches treated as leprosy

16 years, boy

Stage IB

CD8+

A 16-year-old student from a village near Nellore has pale, slightly scaly patches on the trunk, buttocks and thighs for 3 years. He received a course of leprosy treatment and antifungal creams; the patches kept spreading. No itch.

*Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?*

1

#### SUSPECT

Hypopigmented patches in covered skin that change in size and do not respond to treatment for leprosy or fungal infection: hypopigmented MF is in the differential.

2

#### BIOPSY

Creams stopped for 3 weeks; two 4 mm punches: epidermotropic atypical lymphocytes without spongiosis, CD8+, CD7 loss; clonal TCR. ISCL score 5.

3

#### STAGE

Patches cover about 15% of the skin; no nodes; blood flow cytometry BO: stage IB (T2a NO MO BO). Chest X-ray only (CT is for T2b and above).

4

#### TREAT

NBUVB phototherapy (or topical steroid for small areas); slow taper after clearing. Hypopigmented MF responds well and has a favourable outlook.

#### TEACHING POINTS

- Hypopigmented MF is the commonest form in children and darker skin.
- Mimics: leprosy, vitiligo, tinea versicolor, pityriasis alba.
- Biopsy before more empirical treatment.

**See:** Sections 02, 03 and 06, Algorithm 2.

### CASE 2 Classic MF · stage IB

#### Six years of "eczema" on the buttocks

52 years, woman

Stage IB

Patches and thin plaques

A 52-year-old teacher has itchy, scaly, reddish patches on the buttocks, hips and breasts for 6 years, labelled eczema and psoriasis. They cover about 20% of her skin.

*Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?*

1

#### DIAGNOSE

Topicals stopped for 2–4 weeks; biopsy from the thickest plaque: epidermotropism, Pautrier microabscesses, CD4+ with CD7 loss; clone present. MF.

2

#### STAGE

T2b (plaques  $\geq 10\%$ ), no nodes, BO: stage IB. CT or PET-CT for T2b; normal.

3

**TREAT**

Skin-directed: NBUVB or PUVA (or mechlorethamine and potent steroid); clears in months; slow maintenance taper.

4

**FOLLOW**

Relapse after stopping maintenance is likely and usually responds again; rotate skin-directed options; systemic agents only if control fails.

**TEACHING POINTS**

- Persistent patches in sun-protected skin: biopsy.
- Stage IIB MF: skin-directed treatment, not chemotherapy.
- Most patients with patches never progress.

**See:** Sections 03, 05 and 06, Algorithm 2.

**CASE 3 MF · tumours with LCT****New nodules in old plaques**

64 years, man

Stage IIB

CD30+ LCT

A 64-year-old with plaque MF for 8 years develops several fast-growing nodules on the trunk and thigh, two of them ulcerated.

*Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?*

1

**REBIOPSY**

Nodule biopsy: large cells over 25% of the infiltrate (large cell transformation), CD30 in 60%.

2

**STAGE**

PET-CT: no nodes over 1.5 cm, no viscera; blood BO: stage IIB with LCT. CLIP: age >60, LCT → 2 points.

3

**TREAT**

Brentuximab vedotin (ALCANZA) with local RT 8–12 Gy to the ulcerated tumours; watch for neuropathy.

4

**PLAN**

Fit and 64: refer for allogeneic transplant assessment, to be done at best response; mogamulizumab avoided close to transplant.

**TEACHING POINTS**

- A change of pace means a new biopsy.
- LCT and generalised tumours: BV.
- Allogeneic HCT is the only cure in advanced MF.

**See:** Sections 04 and 06, Algorithm 3.

**CASE 4 Sézary syndrome****Red, peeling skin all over and unbearable itch**

68 years, man

Stage IVA1

B2

A 68-year-old retired engineer has 4 months of red, scaly skin covering almost his whole body, hair loss, thick palms and soles, and itch that stops him sleeping. Treated for a drug rash. Firm nodes in both groins.

*Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?*

1

**DIAGNOSE**

Skin biopsy compatible with MF/SS; flow cytometry: 3200/μL CD4+CD7- cells; the same TCR clone in blood and skin. HTLV-1 negative. Sézary syndrome.

2

**STAGE**

Groin node excision: architecture preserved (N2); PET-CT: no viscera: T4 N2 M0 B2 = stage IVA1.

3

**TREAT**

Mogamulizumab, or ECP with interferon at a specialised centre; add bexarotene or interferon if needed.

4

**SUPPORT**

Emollients, bedtime doxepin, gabapentin; skin swabs; bleach baths and topical antibiotic for colonisation; prompt antivirals for herpes.

**TEACHING POINTS**

- Erythroderma: flow cytometry before calling it a drug rash.
- Sézary: ECP or mogamulizumab first.
- Treat itch and infection as seriously as the lymphoma.

**See:** Sections O3 and O6 (Sézary), Algorithm 3.

**CASE 5** Lymphomatoid papulosis**Crops of papules that crust and heal**

34 years, woman

LyP type A

CD30+

A 34-year-old nurse has crops of red papules on the arms and thighs for 2 years; each crusts in the centre and heals in 4–6 weeks with small scars. She feels well.

*Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?*

1

**DIAGNOSE**

Excision of two fresh papules: wedge-shaped mixed infiltrate with large CD30+ cells, ALK negative; TCR clone. LyP type A.

2

**TREAT**

Few lesions: observation or a super-potent topical steroid. If they become many or appear on her face or hands: low-dose weekly methotrexate (liver and blood monitoring), then PUVA.

3

**WATCH**

A 3 cm nodule that has not regressed in 8 weeks: excise to look for primary cutaneous ALCL (CD30 on ≥75% of large cells); ALK, HTLV-1 and staging if so.

4

**FOLLOW**

Lifelong review: about 15% develop another lymphoma (MF, ALCL). No chemotherapy for LyP itself.

**TEACHING POINTS**

- Self-healing CD30+ papules: LyP, not high-grade lymphoma.
- Treatment is for symptoms; it does not change the course.
- Biopsy persistent nodules over 2 cm.

**See:** Section O6 (CD30+), Algorithm 4.

**CASE 6** Primary cutaneous ALCL · localised**An ulcerating nodule on the forearm**

59 years, man

Solitary

CD30 ≥75%

A 59-year-old farmer has a 3 cm red nodule on the forearm that grew over 2 months and ulcerated. Antibiotics for a presumed abscess did not help. No fever or weight loss; a soft 1.5 cm node in the axilla.

*Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?*

**1** **DIAGNOSE**

Excision biopsy: sheets of large anaplastic cells, CD30 on over 90%, CD4+, ALK and EMA negative; HTLV-1 negative. Primary cutaneous ALCL.

**2** **STAGE**

Axillary node biopsy: reactive (dermatopathic) only. PET-CT: no other disease. Localised skin disease, no nodal involvement.

**3** **TREAT**

The excision has clear margins: observation; adding RT after excision gives no benefit. Had excision not been possible: involved-site RT (low doses, ≤20 Gy, appear enough).

**4** **FOLLOW**

Skin relapse is common (about 40%) but rarely dangerous: a new nodule is observed first, then excised or irradiated; brentuximab vedotin is kept for multifocal disease with nodes or symptoms, or multiple relapses.

**TEACHING POINTS**

- An ulcerating nodule that is not an abscess: biopsy.
- Localised C-ALCL: excision or RT, never CHOP.
- Biopsy enlarged nodes: most are reactive.

**See:** Section 06 (CD30+), Algorithm 4.

**CASE 7** PCFCL · scalp**A slowly growing scalp nodule**

55 years, man

T1a

BCL6+ BCL2-

A 55-year-old shop owner has a firm, painless 3 cm pink nodule on the scalp that has grown over a year. No fevers or weight loss.

*Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?*

**1** **DIAGNOSE**

Excision biopsy: dermal infiltrate sparing the epidermis, large centrocytes; CD20+, BCL6+, BCL2-, MUM1-; FOXP1 and IgM not strong. PCFCL.

**2** **STAGE**

PET-CT: no other disease; CBC and LDH normal: T1a NO MO.

**3** **TREAT**

RT to the lesion with a 1–1.5 cm margin (response-adapted 4 Gy, escalating to 24 Gy; higher doses give better local control).

**4** **FOLLOW**

Every 3 months for 2 years with skin examination; no routine scans. A skin relapse outside the field → RT again.

**TEACHING POINTS**

- PCFCL is indolent: 5-year survival about 95%.
- BCL2, MUM1, FOXP1, IgM strong → leg-type DLBCL instead.
- Exclude systemic follicular lymphoma with PET-CT.

**See:** Sections 05 and 06 (B-cell), Algorithm 5.

**CASE 8 SPTCL · HLH****Fat nodules, fever and falling counts**

28 years, woman

Lupus

SPTCL

A 28-year-old with lupus has painless nodules on her thighs and arms for 7 months, called lupus panniculitis. She now has high fevers, a large liver, falling counts and a very high ferritin.

*Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?*

1

**SUSPECT**

Painless nodules that come and go, then fever and cytopenias: SPTCL with HLH must be excluded (lupus profundus nodules are usually painful).

2

**BIOPSY**

Deep excisional biopsy with fat: atypical lymphocytes rimming fat cells; CD3+, CD8+,  $\beta$ F1+, cytotoxic; CD4-, CD56-; EBER negative; clonal TCR. SPTCL.

3

**URGENT**

HLH work-up (ferritin, triglycerides, fibrinogen) and urgent haematology care; HLH is the main factor for poor outcome (5-year survival 46% vs 91%).

4

**PLAN**

Treatment is planned with the lymphoma team (SPTCL treatment is not yet covered in this module). Germline HAVCR2 (TIM-3) variants are common in SPTCL with HLH.

**TEACHING POINTS**

- Shave biopsies miss SPTCL: biopsy the fat.
- Up to 20% of SPTCL patients have autoimmune disease.
- Fever and cytopenias in SPTCL: think HLH.

**See:** Sections 02, 03 and 06 (rare subtypes), Algorithm 4.

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- **Sézary syndrome at Mayo (Kubica 2012):** Kubica AW, Davis MD, Weaver AL, et al. Sézary syndrome: a study of 176 patients at Mayo Clinic. *J Am Acad Dermatol* 2012;67:1189. [PubMed](#) Classification, diagnosis, staging and prognosis
- **4 Gy RT for indolent B-cell lymphoma (Imber 2021):** Imber BS, Chau KW, Lee J, et al. Excellent response to very-low-dose radiation (4 Gy) for indolent B-cell lymphomas. *Blood Adv* 2021;5:4185. [PubMed](#) Cutaneous B-cell lymphomas and follow-up

Educational summary for health professionals, written in our own words from current guidelines and primary trials. No drug doses are given. Hodgkin, aggressive B-cell, indolent B-cell and T-cell lymphomas are parts 1–4; primary cutaneous diffuse large B-cell lymphoma, leg type, is

covered in the aggressive B-cell module. Verify current guidelines, Indian regulatory approvals and individual patient factors before treatment decisions.