

Sarcoma

102 questions in three levels, with answer key

For NEET-PG, USMLE, DNB and DM revision and for classroom tests. Level 1 covers core facts; Level 2 tests applied reasoning (calculations, imaging and fluid findings, trial interpretation, sequencing); Level 3 covers landmark trials and newer therapies. Answers and explanations are at the end. No drug doses.

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Level 1 · Basics

- Q1. Soft tissue** Which statement about the relative frequency of sarcomas is correct?
- A Soft tissue sarcomas outnumber bone sarcomas by roughly four to one
 - B Osteosarcoma is the commonest sarcoma in older adults
 - C Soft tissue and bone sarcomas are about equally common
 - D Sarcomas account for about 10% of adult cancers
 - E Bone sarcomas outnumber soft tissue sarcomas by roughly four to one
- Q2. Soft tissue** Which three types are the commonest soft tissue sarcomas in adults?
- A Rhabdomyosarcoma, synovial sarcoma and Ewing sarcoma
 - B Angiosarcoma, Kaposi sarcoma and MPNST
 - C Alveolar soft part, clear cell and epithelioid sarcoma
 - D Desmoid tumour, GIST and dermatofibrosarcoma protuberans
 - E Undifferentiated pleomorphic sarcoma, liposarcoma and leiomyosarcoma
- Q3. Bone** A 15-year-old boy has had knee pain for 3 months that now wakes him at night, with a firm swelling above the knee. Which tumour is most likely?
- A Conventional chondrosarcoma
 - B Osteochondroma
 - C Osteosarcoma of the distal femur
 - D Synovial sarcoma of the knee joint
 - E Chronic osteomyelitis
- Q4. Bone** This schematic radiograph is from a 15-year-old with night pain above the knee. Which finding points to osteosarcoma?
- A Multiple well-defined lucent lesions in the epiphysis
 - B A uniformly sclerotic bone with no soft tissue mass
 - C A calcified cartilage cap on a bony stalk
 - D A sunburst periosteal reaction with a Codman triangle in the metaphysis
 - E Onion-skin layering of the diaphysis
- Q5. Bone** Where is Ewing sarcoma most characteristically found?
- A In the epiphysis of the proximal tibia in older adults
 - B In the metaphysis of the distal femur in teenagers only
 - C In the diaphysis of a long bone, the pelvis or the ribs, in children and young adults
 - D In the phalanges of the hand in middle age
 - E In the skull vault in adults over 60
- Q6. Bone** Which statement about conventional chondrosarcoma is correct?
- A It mostly affects children under 10 and responds well to chemotherapy
 - B It mainly affects adults over 40 and is chemoresistant, so surgery is the only effective treatment
 - C It is treated with the same chemotherapy as osteosarcoma
 - D It is radiosensitive and is managed with radiotherapy alone
 - E It never occurs in the pelvis
- Q7. Soft tissue** A 45-year-old has a painless, slowly enlarging 7 cm lump deep in the thigh. What is the most appropriate next step?
- A Observation for 3 months
 - B FNAC alone, with reassurance if it is benign
 - C Ultrasound only, and discharge if there is no pain
 - D MRI of the thigh with contrast before any biopsy or excision
 - E Excision biopsy under local anaesthesia

- Q8. Soft tissue** Which feature most raises the suspicion that a soft tissue lump is a sarcoma?
- A A history of a single minor injury
 - B Pain only at night
 - C A soft consistency
 - D Tenderness on palpation
 - E Size over 5 cm, depth below the fascia, or growth
- Q9. Soft tissue** Which biopsy approach is preferred for a suspected limb sarcoma?
- A Frozen section during a planned excision elsewhere
 - B Excision of the whole lump as the biopsy
 - C A transverse incisional biopsy
 - D Image-guided core needle biopsy after MRI, along a track that will be removed with the tumour
 - E FNAC alone
- Q10. Soft tissue** Which grading system is used for soft tissue sarcomas?
- A WHO CNS grading
 - B Nottingham (Bloom–Richardson)
 - C Fuhrman
 - D Gleason
 - E FNCLCC (differentiation, mitotic count and necrosis)
- Q11. Soft tissue** Which investigation should every adult with a new soft tissue sarcoma have to look for metastases?
- A CT of the chest
 - B MRI of the brain
 - C Serum LDH
 - D Chest radiograph only
 - E Bone scan
- Q12. Soft tissue** Which fusion characterises synovial sarcoma?
- A EWSR1::FLI1
 - B COL1A1::PDGFB
 - C FUS::DDIT3
 - D PAX3::FOXO1
 - E SS18::SSX1/2/4
- Q13. Soft tissue** MDM2 amplification on FISH helps to distinguish which pair?
- A GIST from desmoid tumour
 - B Synovial sarcoma from Ewing sarcoma
 - C Osteosarcoma from osteomyelitis
 - D Leiomyosarcoma from leiomyoma
 - E Well-differentiated liposarcoma from lipoma
- Q14. Soft tissue** A 28-year-old woman has an osteosarcoma. Her mother had breast cancer at 36 and her brother had an adrenocortical tumour in childhood. Which germline gene should be tested?
- A VHL
 - B TP53
 - C APC
 - D MLH1
 - E NF2
- Q15. Soft tissue** A patient with neurofibromatosis type 1 has a plexiform neurofibroma that has recently become painful and hard. What is the concern and the next step?
- A Normal growth of a neurofibroma; reassure
 - B A lipoma; excise locally
 - C A schwannoma; observe
 - D A desmoid tumour; start imatinib
 - E Malignant peripheral nerve sheath tumour; MRI with or without PET, then biopsy

- Q16. Soft tissue** What is the usual latency of a radiation-associated sarcoma?
- A About 1–2 years
 - B Under 6 months
 - C Usually 7–16 years, shorter (4–8 years) for angiosarcoma after breast radiotherapy
 - D Always over 40 years
 - E There is no latency; they appear during radiotherapy
- Q17. Soft tissue** A long-standing swollen arm after mastectomy and axillary surgery develops purple nodules. Which diagnosis must be excluded by biopsy?
- A Lymphangiosarcoma (Stewart–Treves syndrome)
 - B Lipoedema
 - C Venous ulceration
 - D Contact dermatitis
 - E Recurrent cellulitis
- Q18. Soft tissue** What did the NCI trial by Rosenberg (1982) show?
- A Radiotherapy alone was as good as surgery
 - B Amputation gave better survival than limb salvage
 - C Limb-sparing surgery with radiotherapy gave the same survival as amputation
 - D Preoperative radiotherapy reduced distant metastases
 - E Adjuvant chemotherapy cured most patients
- Q19. Soft tissue** A “lipoma” is excised in a minor theatre and histology shows a high-grade sarcoma with involved margins. What should be done next?
- A MRI of the site and re-excision of the scar and tumour bed, after expert pathology review
 - B No further local treatment, because the lump has been removed
 - C Reassure and review in 3 months
 - D Postoperative chemotherapy alone
 - E Wait for a local recurrence
- Q20. Soft tissue** What is the standard preoperative radiotherapy dose for a soft tissue sarcoma of the limb?
- A 50 Gy in 25 fractions
 - B 66 Gy in 33 fractions
 - C 20 Gy in 5 fractions
 - D 30 Gy in 10 fractions
 - E 75 Gy in 30 fractions
- Q21. Soft tissue** Compared with postoperative radiotherapy, preoperative radiotherapy for limb sarcoma causes:
- A Worse local control
 - B Worse overall survival
 - C More late fibrosis and joint stiffness
 - D No wound complications
 - E More wound complications but less permanent late fibrosis
- Q22. Soft tissue** Which statement about retroperitoneal sarcoma is correct?
- A It is usually under 3 cm at diagnosis
 - B It is cured by biopsy and radiotherapy alone
 - C It nearly always presents with early jaundice
 - D It commonly spreads to lymph nodes
 - E It is often silent until large, about 15 cm at diagnosis
- Q23. Bone** What does MAP stand for in osteosarcoma chemotherapy?
- A Melphalan, doxorubicin and carboplatin
 - B Mesna, ifosfamide and pazopanib
 - C Methotrexate, actinomycin and procarbazine
 - D Mitomycin, ifosfamide and paclitaxel
 - E High-dose methotrexate, doxorubicin (Adriamycin) and cisplatin

- Q24. Bone** Which regimen is the backbone of Ewing sarcoma chemotherapy?
- A MAP
 - B VDC/IE (vincristine, doxorubicin, cyclophosphamide alternating with ifosfamide and etoposide)
 - C Doxorubicin plus trabectedin
 - D Gemcitabine plus docetaxel
 - E Weekly paclitaxel
- Q25. Bone** Which factor is the strongest prognostic marker after metastatic status in osteosarcoma?
- A Histological necrosis in the resection specimen after preoperative chemotherapy (90% or more is a good response)
 - B How much the mass shrinks on X-ray
 - C PD-L1 expression
 - D Serum CEA
 - E The patient's height
- Q26. Metastatic** What is the reference first-line drug for most patients with metastatic soft tissue sarcoma?
- A Gemcitabine alone
 - B Doxorubicin
 - C Pazopanib
 - D Cisplatin
 - E Imatinib
- Q27. Soft tissue** Which drug requires an echocardiogram before treatment and a record of the lifetime cumulative dose?
- A Doxorubicin
 - B Eribulin
 - C Trabectedin
 - D Methotrexate
 - E Pazopanib
- Q28. Soft tissue** Which measure prevents ifosfamide haemorrhagic cystitis?
- A Dexrazoxane
 - B Mesna with hydration
 - C Leucovorin
 - D Allopurinol
 - E Amifostine
- Q29. Metastatic** In advanced soft tissue sarcoma, pazopanib is not used for which group?
- A MPNST
 - B Synovial sarcoma
 - C Leiomyosarcoma
 - D Adipocytic sarcomas (liposarcoma)
 - E Undifferentiated pleomorphic sarcoma
- Q30. Soft tissue** Which drug is the usual first line for cutaneous angiosarcoma of the scalp and face?
- A Tazemetostat
 - B High-dose methotrexate
 - C Imatinib
 - D Weekly paclitaxel
 - E Pazopanib
- Q31. Soft tissue** When do most relapses of soft tissue sarcoma occur?
- A Relapse is rare after complete resection
 - B In the first 2–3 years, so surveillance is front-loaded
 - C Only in the first month
 - D Evenly across 10 years
 - E After 15 years

Q32. Bone Before cisplatin, ifosfamide or other alkylating chemotherapy in a 15-year-old boy, which step is often forgotten?

- A Thyroid screening
- B A live vaccine
- C Sperm banking
- D Dental extraction
- E Bone marrow harvest

Q33. Bone Which investigation should come first in a teenager with persistent bone pain?

- A Serum calcium only
- B Bone biopsy at the local hospital
- C A plain X-ray of the whole bone in two planes
- D A bone density scan
- E A trial of antituberculous treatment

Q34. Soft tissue Which statement about lymph node metastasis in sarcoma is correct?

- A Nodes are involved in most soft tissue sarcomas
- B Nodal spread is the commonest route of metastasis
- C Nodal disease never occurs in sarcoma
- D Sentinel node biopsy is routine in all sarcomas
- E Nodal involvement is uncommon (under 5%) except in clear cell, epithelioid, angiosarcoma and rhabdomyosarcoma

Level 2 · Applied & innovative

Q35. Soft tissue A 52-year-old man had an 8 cm "lipoma" of the thigh excised under local anaesthesia, with a drain brought out 10 cm away. Histology: high-grade pleomorphic sarcoma with involved margins. Which plan is most appropriate?

- A Adjuvant chemotherapy alone
- B MRI of the thigh and CT chest, expert pathology review, preoperative RT that includes the scar and drain track, then re-excision
- C Hip disarticulation
- D Observation with MRI at 6 months
- E Re-excise only the scar, without radiotherapy

Q36. Soft tissue In SU2C-SARCO32 (stage III limb UPS or pleomorphic or dedifferentiated liposarcoma), adding pembrolizumab to preoperative RT and surgery:

- A Had no effect on disease-free survival
- B Improved 2-year disease-free survival (67 vs 52%) with more grade 3 or worse toxicity (56 vs 31%); overall survival is not yet known
- C Was tested together with doxorubicin-ifosfamide chemotherapy
- D Improved overall survival by 20% with no extra toxicity
- E Replaced the need for surgery

Q37. Soft tissue Which patient with a limb soft tissue sarcoma can reasonably be treated with wide excision alone?

- A A deep 9 cm grade 3 tumour
- B Any tumour next to a nerve
- C A tumour with a positive margin that cannot be re-excised
- D An 8 cm myxofibrosarcoma with infiltrative margins
- E A superficial 4 cm grade 1 tumour with clear margins

Q38. Soft tissue After preoperative RT, a thigh sarcoma is resected with a 2 mm margin where it abuts an intact fascial plane, and the sciatic nerve is preserved. How is this margin regarded?

- A Unacceptable unless a 5 cm margin is achieved
- B Often adequate: fascia, periosteum and vessel adventitia are good barriers, and planned close margins beside a preserved nerve or vessel are acceptable after preoperative RT
- C Always an R2 resection
- D An indication for amputation
- E Impossible to judge without a PET scan

- Q39. Soft tissue** What target volume was used for preoperative RT in RTOG 0630?
- A The tumour only, with no margin
 - B The tumour plus 10 cm in every direction
 - C The regional lymph node basin
 - D The tumour plus about 1.5 cm radially and 3–4 cm along the muscle, including T2 oedema where feasible
 - E The whole limb
- Q40. Soft tissue** A 34-year-old man has this deep posterior thigh mass (schematic T2 MRI). Core biopsy: myxoid liposarcoma with a FUS::DDIT3 fusion. Which staging is specific to this subtype?
- A PET-CT alone
 - B Bone marrow aspirate only
 - C CT chest only
 - D CT chest, CT abdomen–pelvis and whole–spine MRI; PET may miss disease
 - E MRI of the brain
- Q41. Soft tissue** In the DOREMY phase 2 trial of preoperative RT for myxoid liposarcoma, 36 Gy gave:
- A A higher amputation rate
 - B Worse local control than surgery alone
 - C No pathological response
 - D 91% extensive pathological response and no local failures
 - E Results proven equal to 50 Gy in a phase 3 trial
- Q42. Soft tissue** A 58-year-old woman has abdominal fullness. This schematic CT shows a large fatty left retroperitoneal mass with thick septa and a solid enhancing nodule. What does the nodule suggest, and what is the best next step?
- A A benign lipoma; observe
 - B A germ cell tumour; orchidectomy
 - C Lymphoma; excision biopsy at laparotomy
 - D Renal angiomyolipoma; embolise
 - E Dedifferentiation within a well-differentiated liposarcoma; percutaneous posterior core biopsy of the nodule and discussion at a sarcoma MDT
- Q43. Soft tissue** How should the result of STRASS be applied to retroperitoneal sarcoma?
- A Preoperative RT is standard for every retroperitoneal sarcoma
 - B Preoperative RT did not improve abdominal recurrence-free survival overall; it is an option in selected well-differentiated and low-grade dedifferentiated liposarcoma after MDT discussion
 - C Radiotherapy should never be used
 - D Chemotherapy replaces surgery
 - E Postoperative RT is preferred
- Q44. Soft tissue** At the first operation for a resectable retroperitoneal liposarcoma, which approach gives the best local control?
- A Compartmental en bloc resection including adherent organs, by an experienced team
 - B Enucleation along the capsule
 - C Laparoscopic morcellation
 - D Debulking with planned reoperation
 - E Open biopsy followed by chemotherapy
- Q45. Soft tissue** A 47-year-old woman had a hysterectomy for a rapidly growing “fibroid” that proved to be leiomyosarcoma, and now has a cough. Which staging is most appropriate?
- A Bone scan only
 - B MRI of the brain
 - C Chest X-ray only
 - D No imaging, because the uterus has been removed
 - E CT of the chest and abdomen including the liver

- Q46. Soft tissue** A 64-year-old woman treated with breast-conserving surgery and RT 7 years ago has had this violaceous, bruise-like patch for 2 months and was given antibiotics for "cellulitis". What is the next step?
- A Topical steroids
 - B A longer course of antibiotics
 - C Observation for 6 months
 - D Multiple punch biopsies from the darkest areas
 - E Mammography alone and reassurance
- Q47. Soft tissue** Biopsy confirms radiation-associated angiosarcoma of the breast. Which statement about local treatment is correct?
- A Chemotherapy replaces surgery for localised disease
 - B Imatinib is first line
 - C Radical resection of all irradiated skin where possible, with reconstruction, is the main treatment; the disease is often wider than imaging suggests
 - D Radiotherapy alone is curative
 - E Wide local excision with a 1 cm margin is sufficient
- Q48. Soft tissue** After breast-conserving therapy the 10-year incidence of sarcoma is about 0.16% (Wiener 2023). What follows from this?
- A It is not a reason to withhold breast radiotherapy when it is indicated
 - B Radiotherapy should be avoided in all breast cancer
 - C Angiosarcoma is unrelated to radiotherapy
 - D Every survivor needs yearly breast skin MRI
 - E Sarcoma is commoner than breast cancer recurrence
- Q49. Bone** A 15-year-old's X-ray shows a sunburst lesion of the distal femur. Which sequence is correct?
- A Immediate biopsy at the nearest hospital, then referral
 - B MRI of the whole femur, CT chest and PET-CT or bone scan, then core biopsy at the centre that will operate
 - C Chemotherapy before any tissue diagnosis
 - D Amputation without biopsy
 - E Curettage as for a suspected infection
- Q50. Bone** A 24-year-old man has 4 months of thigh pain, fever and a raised ESR, and antituberculous treatment was started for a "lytic lesion" without a biopsy. His radiograph (schematic) shows a permeative, moth-eaten pattern with onion-skin layering and a soft tissue mass. What should be done?
- A Six weeks of antibiotics without a biopsy
 - B CT-guided core biopsy sent for histology, molecular testing and culture, and stop the unproven antituberculous treatment
 - C Curettage and cementing
 - D Bone scan only
 - E Continue antituberculous drugs for 6 months
- Q51. Bone** A 52-year-old has a high-grade osteosarcoma. Which chemotherapy is usual?
- A Pazopanib
 - B VDC/IE
 - C No chemotherapy, because adults do not benefit
 - D MAP exactly as for a teenager
 - E Doxorubicin-cisplatin; the benefit of high-dose methotrexate is unproven at this age
- Q52. Bone** A teenager's osteosarcoma shows 60% necrosis after preoperative MAP. What should the postoperative chemotherapy be?
- A Add ifosfamide-etoposide, which improved survival
 - B Switch to VDC/IE
 - C Continue MAP to complete about 29 weeks; changing or intensifying treatment did not help in EURAMOS-1
 - D Stop chemotherapy
 - E Add zoledronate to improve survival

- Q53. Bone** Which is the correct overall sequence for localised Ewing sarcoma in a 17-year-old?
- A VDC/IE only after surgery
 - B VDC/IE every 14 days for 12 weeks, local control (surgery or RT), then chemotherapy to a total of 14–17 cycles
 - C Surgery first, then 6 cycles of MAP
 - D Interferon maintenance after 4 cycles
 - E Radiotherapy alone
- Q54. Bone** What did AEWS0031 show?
- A Adding interferon improved survival
 - B Cycles every 14 days added toxicity with no benefit
 - C VDC/IE every 14 days instead of 21 improved 10-year event-free survival (70 vs 61%) without more toxicity
 - D Adding vincristine–topotecan–cyclophosphamide improved survival
 - E A higher dose per cycle improved survival
- Q55. Bone** A Ewing sarcoma of the iliac wing with a large mass is judged unresectable without mutilation. Which local treatment is appropriate?
- A Radiotherapy after 3 years
 - B Definitive RT (about 55 Gy) with chemotherapy continuing, omitting doxorubicin during RT
 - C Hemipelvectomy regardless of function
 - D No local treatment; chemotherapy alone
 - E Intralesional curettage
- Q56. Metastatic** A fit 47-year-old woman has metastatic uterine leiomyosarcoma (lung and liver). Which first-line regimen improved both PFS and overall survival in a randomised trial?
- A Gemcitabine–docetaxel
 - B Doxorubicin plus trabectedin, followed by trabectedin maintenance (LMS-04)
 - C Pazopanib alone
 - D Imatinib
 - E Observation
- Q57. Metastatic** In EORTC 62012, adding ifosfamide to doxorubicin in first-line metastatic soft tissue sarcoma:
- A Doubled the response rate (26 vs 14%) and lengthened PFS without a significant survival gain
 - B Reduced toxicity
 - C Was inferior in every subtype
 - D Had no effect on response
 - E Improved overall survival significantly
- Q58. Metastatic** A 68-year-old has metastatic dedifferentiated liposarcoma and wants symptom control with the least toxicity. What is the first line?
- A Doxorubicin plus ifosfamide at once
 - B Pazopanib
 - C Imatinib
 - D Trabectedin alone in every case
 - E Single-agent doxorubicin; combinations only when a response is needed
- Q59. Metastatic** A patient with advanced PEComa should receive:
- A An mTOR inhibitor (nab-sirolimus; sirolimus or everolimus where it is unavailable)
 - B Cisplatin
 - C Doxorubicin plus ifosfamide
 - D Weekly paclitaxel
 - E Imatinib
- Q60. Metastatic** A young adult with an advanced sarcoma has an NTRK fusion found by RNA sequencing. Which treatment gives high, durable responses?
- A Pazopanib
 - B Mifamurtide
 - C Imatinib
 - D Larotrectinib or entrectinib
 - E Atezolizumab

- Q61. Metastatic** Three years after treatment of a grade 3 synovial sarcoma near the knee, a 29-year-old woman has two new right-lung nodules on surveillance CT (schematic), with nothing elsewhere on PET-CT. What is the best approach?
- A Adjuvant MAP after resection
 - B Observe until symptoms develop
 - C Whole-lung radiotherapy
 - D Repeat CT at 6–8 weeks to test the tempo, then VATS resection if no new lesions, with no routine adjuvant chemotherapy
 - E Start palliative chemotherapy at once
- Q62. Bone** Which medicines should be avoided during high-dose methotrexate for osteosarcoma, and what is used for severely delayed clearance?
- A Avoid NSAIDs, PPIs and co-trimoxazole; glucarpidase for delayed clearance
 - B Avoid allopurinol; give hydroxocobalamin
 - C Avoid statins; give dexrazoxane
 - D Avoid mesna; give amifostine
 - E Avoid antiemetics; give rasburicase
- Q63. Bone** Which long-term effects should be anticipated after cisplatin and ifosfamide for a bone sarcoma?
- A Pulmonary fibrosis and hypothyroidism only
 - B Cardiomyopathy from cisplatin
 - C Hair depigmentation
 - D Peripheral neuropathy only from ifosfamide
 - E Hearing loss and renal tubular loss of phosphate and magnesium
- Q64. Soft tissue** For how long should retroperitoneal liposarcoma be followed with imaging?
- A For 6 months
 - B Only if symptoms develop
 - C For 2 years
 - D Beyond 10 years, because liposarcoma recurs late and can dedifferentiate
 - E Until the first scan is clear
- Q65. Soft tissue** Which imaging is preferred to follow the primary site after limb sarcoma resection with positive margins or at a hard-to-examine site?
- A No imaging
 - B PET-CT every 3 months
 - C Ultrasound every 3 months only
 - D MRI about 6-monthly for 2 years, then yearly to 5 years
 - E Chest radiograph yearly
- Q66. Bone** Which statement about late effects after treatment of a bone sarcoma in a young person is correct?
- A Survivors cannot play sport
 - B An echocardiogram is needed only at diagnosis
 - C Anthracycline cardiomyopathy, cisplatin hearing loss, infertility and second cancers can appear years later and need survivorship follow-up
 - D Late effects do not occur after chemotherapy
 - E Only radiotherapy causes late effects
- Q67. Soft tissue** Which statement about GIST and desmoid tumour is correct?
- A GIST responds to MAP
 - B Both are bone tumours
 - C Desmoid tumour is always treated by amputation
 - D They are separate diseases, not treated like soft tissue sarcoma: imatinib for GIST, and initial observation for desmoid
 - E Both are treated with doxorubicin–ifosfamide

- Q68. Bone** Why is preoperative chemotherapy preferred in osteosarcoma?
- A It gives better survival than postoperative chemotherapy
 - B It prevents all metastases
 - C It allows radiotherapy to be omitted
 - D It eases limb salvage and allows necrosis to be assessed; survival is similar to postoperative timing
 - E It removes the need for surgery

Level 3 · Challenging

- Q69. Bone** What did the Multi-Institutional Osteosarcoma Study (MIOS) establish?
- A Adjuvant chemotherapy after surgery raised relapse-free survival from about 17% to about 65%
 - B Interferon maintenance prolonged survival
 - C Limb salvage was inferior to amputation
 - D Radiotherapy can replace surgery
 - E Zoledronate improved survival
- Q70. Bone** In INT-0091, adding ifosfamide and etoposide to VDC in localised Ewing sarcoma gave 5-year event-free survival of:
- A 45% versus 44%
 - B 30% versus 20%
 - C 54% versus 69%
 - D 69% versus 54%
 - E 90% versus 70%
- Q71. Bone** In Euro Ewing 2012, VDC/IE compared with VIDE gave:
- A 3-year EFS 67 vs 61% and overall survival 82 vs 74%, with less febrile neutropenia
 - B More toxicity with VDC/IE
 - C Worse survival with VDC/IE
 - D Better results only in adults
 - E No difference in any outcome
- Q72. Soft tissue** How should the pooled evidence for adjuvant doxorubicin-ifosfamide (Pervaiz 2008, EORTC 62931) be applied?
- A Give it to every patient with soft tissue sarcoma
 - B Give it only to patients over 70
 - C Give it to all low-grade tumours
 - D Never give it
 - E Offer chemotherapy only to fit patients with high-risk tumours (over 5 cm, deep, grade 2–3, ideally chemosensitive types) after shared decision-making
- Q73. Soft tissue** ISG-STS 1001 compared three cycles of epirubicin plus ifosfamide with histotype-tailored regimens in high-risk soft tissue sarcoma. What was the conclusion?
- A Five cycles were superior to three
 - B The standard regimen was better overall; high-grade myxoid liposarcoma, where the tailored drug is trabectedin, was the exception
 - C The trial was negative for toxicity only
 - D Tailored regimens were superior in every histotype
 - E Both arms had identical results in every histotype
- Q74. Metastatic** Which statement about first-line trials in metastatic soft tissue sarcoma is correct?
- A Gemcitabine-docetaxel was superior to doxorubicin
 - B Gemcitabine-docetaxel was not better than doxorubicin and was more toxic (GeDDiS); olaratumab's phase 2 signal was not confirmed in phase 3
 - C Olaratumab doubled survival in phase 3
 - D Pazopanib beat doxorubicin first line
 - E Trabectedin beat doxorubicin in every subtype

- Q75. Metastatic** Which soft tissue sarcomas respond best to PD-1 blockade, and is PD-L1 a useful selector?
- A All sarcomas equally; only PD-L1-high tumours respond
 - B Leiomyosarcoma and synovial sarcoma; PD-L1 is the selector
 - C Osteosarcoma only
 - D UPS, dedifferentiated liposarcoma, alveolar soft part sarcoma and cutaneous angiosarcoma; PD-L1 is not a useful selector
 - E Ewing sarcoma only
- Q76. Soft tissue** A 22-year-old woman has alveolar soft part sarcoma (ASPSCR1::TFE3) of the thigh with lung metastases. Which statement is correct?
- A Imatinib is standard
 - B Chemotherapy is effective and curative
 - C Doxorubicin is highly effective and should start at once
 - D It is indolent: limited stable disease can be observed, atezolizumab or pembrolizumab is used if it progresses, and brain MRI is part of staging
 - E It never metastasises to the brain
- Q77. Soft tissue** Which pair is correct for epithelioid sarcoma?
- A NR4A3 fusion and middle-aged men
 - B SMARCB1 (INI1) loss and nodal spread, with tazemetostat where approved
 - C ASPSCR1::TFE3 and brain metastases
 - D EWSR1::ATF1 and BRAF V600E
 - E WWTR1::CAMTA1 and liver involvement
- Q78. Soft tissue** A tumour of the ankle tendons in a young adult is S100 and HMB45 positive but BRAF wild-type, with EWSR1::ATF1. Which is correct?
- A Clear cell sarcoma: melanoma-like but distinct; nodal spread occurs and chemotherapy rarely works, so trials are preferred
 - B Chordoma: observe
 - C Metastatic melanoma: treat with BRAF inhibitors
 - D Epithelioid sarcoma: treat with tazemetostat
 - E Synovial sarcoma: treat with ifosfamide
- Q79. Soft tissue** Which tumour is indolent, occurs in middle-aged men in the thigh and carries an NR4A3 fusion?
- A Extraskeletal myxoid chondrosarcoma
 - B Desmoplastic small round cell tumour
 - C Inflammatory myofibroblastic tumour
 - D Epithelioid haemangioendothelioma
 - E Dedifferentiated liposarcoma
- Q80. Soft tissue** Which fusion is characteristic of epithelioid haemangioendothelioma, a slow, often multifocal tumour of liver, lung and bone?
- A NAB2::STAT6
 - B SS18::SSX2
 - C WWTR1::CAMTA1
 - D EWSR1::WT1
 - E ETV6::NTRK3
- Q81. Soft tissue** An ALK-positive inflammatory myofibroblastic tumour of the abdomen in a young adult is unresectable. Which drug class is used?
- A Larotrectinib
 - B Pazopanib
 - C An ALK inhibitor such as crizotinib
 - D Doxorubicin–ifosfamide first
 - E Imatinib

- Q82. Soft tissue** An adolescent boy has multiple peritoneal masses; biopsy shows small round blue cells with EWSR1::WT1. Which treatment approach is used?
- A MAP
 - B Ewing-type chemotherapy with debulking surgery and whole-abdomen RT in selected patients; the prognosis is poor
 - C Nab-sirolimus
 - D Observation
 - E Imatinib
- Q83. Soft tissue** This schematic H&E field from a pelvic mass in a 19-year-old shows sheets of uniform small round blue cells. Which is the most useful next step?
- A Gleason grading
 - B S100 immunohistochemistry alone
 - C Oestrogen receptor testing
 - D CD99 and NKX2.2 immunohistochemistry with FISH or RNA-based fusion testing, on fresh or frozen tissue
 - E Serum CEA
- Q84. Bone** Which statement about the fusion type in Ewing sarcoma is correct?
- A Fusion testing is unnecessary
 - B The fusion type decides between MAP and VDC/IE
 - C EWSR1::FLI1 (about 85%) and EWSR1::ERG (about 10%) carry the same prognosis with modern therapy
 - D CIC::DUX4 sarcomas are Ewing sarcoma with identical outcomes
 - E EWSR1::ERG has a much worse prognosis
- Q85. Soft tissue** Which are the eligibility requirements for afamitresgene autoleucel in synovial sarcoma, and what is its status in India?
- A HLA-A*02 and MAGE-A4 positivity; not yet available in India, so relevant for trial referral
 - B NTRK fusion; available in India
 - C PD-L1 positivity; widely available
 - D MDM2 amplification; available
 - E BRAF V600E; available
- Q86. Metastatic** Which later-line option is used for indolent dedifferentiated liposarcoma because of CDK4 amplification?
- A Imatinib
 - B Pazopanib
 - C Larotrectinib
 - D A CDK4/6 inhibitor (Dickson 2016)
 - E Tazemetostat
- Q87. Metastatic** In which subtype did eribulin show an overall survival gain in a phase 3 trial (Schöffski 2016)?
- A Liposarcoma
 - B Synovial sarcoma
 - C Angiosarcoma
 - D Osteosarcoma
 - E Leiomyosarcoma
- Q88. Bone** What is the role of IDH1/2 mutation in conventional chondrosarcoma?
- A Diagnostic support in most conventional central tumours; IDH inhibitors are being tested in trials
 - B It selects patients for imatinib
 - C It is absent in chondrosarcoma
 - D It predicts response to MAP
 - E It mandates amputation
- Q89. Bone** Which chemotherapy approach is used for dedifferentiated and for mesenchymal chondrosarcoma?
- A Imatinib for both
 - B Gemcitabine-docetaxel for both
 - C Ewing-type for dedifferentiated, osteosarcoma-type for mesenchymal
 - D None for either
 - E Osteosarcoma-type chemotherapy for dedifferentiated, Ewing-type for mesenchymal

- Q90. Soft tissue** What does the Sarculator nomogram provide, and what level of risk marks “high risk” for a perioperative chemotherapy discussion?
- A Local recurrence only; 5%
 - B A PD-L1 score; 10%
 - C An individual risk estimate for extremity soft tissue sarcoma that adds age, subtype and site to size and grade; a 10-year metastasis risk of about 40% or more
 - D Response to MAP; 90%
 - E Surgical margin width; 1 mm
- Q91. Soft tissue** What is known about tract seeding after a core needle biopsy of a sarcoma?
- A It occurs only after FNAC
 - B It is rare (TARPSWG 2021), and percutaneous posterior core biopsy of a retroperitoneal sarcoma does not raise local recurrence
 - C It is a reason to prefer open biopsy
 - D It occurs in most patients
 - E It is common and a reason to avoid biopsy
- Q92. Soft tissue** What is the status of moderately hypofractionated (15 fractions) or 5-fraction preoperative RT for soft tissue sarcoma?
- A Contraindicated
 - B An established standard that replaces 50 Gy in 25 fractions
 - C Promising control with acceptable wound complication rates, but still investigational (HYPORT-STs, Kalbasi 2020)
 - D Reserved for brachytherapy
 - E Proven inferior
- Q93. Soft tissue** The NCI (Yang 1998) and MSKCC (Pisters 1996) trials of RT after limb-sparing surgery showed:
- A No change in local control
 - B Better overall survival
 - C Better local control without a survival difference
 - D Fewer distant metastases
 - E Higher amputation rates
- Q94. Bone** Which statement about added agents in osteosarcoma is correct?
- A Zoledronate added nothing (OS2006) and interferon maintenance was negative (EURAMOS-1); mifamurtide is used where licensed but is not available in India
 - B Mifamurtide is widely available in India
 - C Ifosfamide–etoposide improved outcomes for poor responders
 - D Zoledronate improved survival
 - E Interferon improved survival for good responders
- Q95. Bone** Which statement about relapsed bone sarcoma is correct?
- A Radiotherapy alone is standard
 - B Relapse is never curable
 - C Only chemotherapy is used
 - D Resect lung metastases where possible; relapse over a year later with a few resectable nodules can still be cured by surgery; options include irinotecan–temozolomide or topotecan–cyclophosphamide in Ewing sarcoma and multitarget TKIs in osteosarcoma
 - E Imatinib is standard
- Q96. Bone** Which statement about metastatic Ewing sarcoma is correct?
- A Whole-lung RT is contraindicated
 - B Metastatic Ewing sarcoma is always cured
 - C Bone marrow involvement carries the best outlook
 - D Surgery alone is standard
 - E Lung-only disease has a better outlook (whole-lung RT after chemotherapy); bone or marrow metastases fare poorly

- Q97. Bone** Which osteosarcoma subtypes usually need surgery alone?
- A Radiation-associated osteosarcoma
 - B High-grade osteosarcoma of the jaw
 - C Low-grade central and parosteal osteosarcoma
 - D Conventional high-grade osteosarcoma
 - E Osteosarcoma in Paget disease
- Q98. Soft tissue** In childhood rhabdomyosarcoma of a parameningeal head and neck site, the standard treatment is:
- A Pazopanib
 - B Radical surgery alone
 - C Chemotherapy with radiotherapy rather than radical surgery
 - D Observation
 - E Wide local excision only
- Q99. Soft tissue** MYC amplification is used to:
- A Diagnose myxoid liposarcoma
 - B Confirm Ewing sarcoma
 - C Separate post-radiation angiosarcoma from an atypical vascular lesion
 - D Select patients for larotrectinib
 - E Choose pazopanib
- Q100. Metastatic** Which monitoring is needed during pazopanib therapy?
- A Audiometry
 - B Weekly CK
 - C Liver tests at 3, 5, 7 and 9 weeks then monthly, blood pressure, and caution with many lung metastases (pneumothorax)
 - D Echocardiogram every cycle only
 - E Daily INR
- Q101. Metastatic** Which monitoring is specific to trabectedin?
- A TSH and cortisol
 - B Echocardiogram and lifetime dose
 - C Audiometry
 - D Urine for blood with mesna
 - E LFTs and CK every cycle, a central line, and dexamethasone premedication
- Q102. Soft tissue** A myxoid liposarcoma of the thigh has been staged with PET-CT only, which was negative. What is the problem?
- A No problem: PET-CT is the best test for this subtype
 - B PET misses myxoid liposarcoma; CT chest, CT abdomen-pelvis and whole-spine MRI are needed
 - C It needs no staging because it is radiosensitive
 - D It needs bone scan only
 - E It needs brain MRI

Answer key

1-A 2-E 3-C 4-D 5-C 6-B 7-D 8-E 9-D 10-E 11-A 12-E 13-E 14-B 15-E 16-C 17-A 18-C 19-A 20-A 21-E 22-E
23-E 24-B 25-A 26-B 27-A 28-B 29-D 30-D 31-B 32-C 33-C 34-E 35-B 36-B 37-E 38-B 39-D 40-D 41-D
42-E 43-B 44-A 45-E 46-D 47-C 48-A 49-B 50-B 51-E 52-C 53-B 54-C 55-B 56-B 57-A 58-E 59-A
60-D 61-D 62-A 63-E 64-D 65-D 66-C 67-D 68-D 69-A 70-D 71-A 72-E 73-B 74-B 75-D 76-D 77-B 78-A
79-A 80-C 81-C 82-B 83-D 84-C 85-A 86-D 87-A 88-A 89-E 90-C 91-B 92-C 93-C 94-A 95-D 96-E
97-C 98-C 99-C 100-C 101-E 102-B

Explanations

Q1 · A There are more than 100 subtypes, and soft tissue sarcoma is about four times commoner than bone sarcoma. Sarcomas are only about 1% of adult cancers but roughly 1 in 7 to 1 in 10 childhood and adolescent cancers.

Q2 · E UPS, liposarcoma and leiomyosarcoma lead in adults. Rhabdomyosarcoma and Ewing sarcoma are mainly tumours of children and young adults, and the others are uncommon. GIST and desmoid are separate diseases.

Q3 · C Osteosarcoma peaks in teenagers and arises in the metaphysis around the knee. Conventional chondrosarcoma is a tumour of adults over 40. Night pain in a teenager is not growing pains.

Q4 · D Bone-forming osteosarcoma gives a mixed lytic and sclerotic metaphyseal lesion with sunburst spicules and a Codman triangle. Onion-skin layering with a permeative diaphyseal pattern suggests Ewing sarcoma, and a cartilage cap suggests an osteochondroma or chondrosarcoma.

Q5 · C Ewing sarcoma arises in long-bone diaphyses, the pelvis and the ribs, and can also arise in soft tissue. It affects children and young adults.

Q6 · B Conventional chondrosarcoma occurs in adults (pelvis, femur, shoulder girdle) and does not respond to chemotherapy. Surgery is the treatment; dedifferentiated and mesenchymal variants are handled differently.

Q7 · D A lump that is large (over 5 cm), deep or growing needs MRI before a knife. Pain does not separate benign from malignant soft tissue masses; size, depth and growth do.

Q8 · E Size, depth and growth are the clues. Pain is uncommon until the tumour is large, and an injury often only draws attention to a tumour that was already present.

Q9 · D Core biopsy with several cores, planned with the surgeon, is the default. FNAC alone is inaccurate for subtype and grade, and a transverse cut contaminates more tissue and enlarges the resection.

Q10 · E FNCLCC grade is built from differentiation, mitoses and necrosis, and predicts metastasis in most subtypes.

Q11 · A The lung holds about 80% of metastases, so CT chest is standard. A chest radiograph is acceptable only for small low-grade tumours, and brain MRI is kept for particular subtypes.

Q12 · E SS18::SSX is the diagnostic fusion of synovial sarcoma. EWSR1::FLI1 is Ewing sarcoma, FUS::DDIT3 myxoid liposarcoma, PAX3::FOXO1 alveolar rhabdomyosarcoma and COL1A1::PDGFB dermatofibrosarcoma protuberans.

Q13 · E MDM2 amplification separates atypical lipomatous tumour or well-differentiated liposarcoma from lipoma, and dedifferentiated liposarcoma from UPS. Any retroperitoneal “UPS” should be tested.

Q14 · B This pattern suggests Li-Fraumeni syndrome (germline TP53): sarcoma at a young age with family cancers. Radiotherapy is avoided where possible in carriers.

Q15 · E MPNST has a lifetime risk of about 8–13% in NF1. Pain, growth or hardening of a plexiform neurofibroma needs MRI, often PET, and biopsy.

Q16 · C The absolute risk is low (well under 1%) but persists for decades. The criteria are a sarcoma in a previously irradiated field, a different histology from the first cancer, and a latency of at least 3 years (classically over 5).

Q17 · A Purple nodules or bruise-like patches on a chronically lymphoedematous limb need a punch biopsy for angiosarcoma (Stewart–Treves).

Q18 · C Limb salvage with radiotherapy became standard, and amputation is now needed in under 5–10% of patients.

Q19 · A Residual tumour is found in about half of unplanned excisions (Noria 1996). Re-image with MRI, review the pathology and re-excise the scar and bed; radiotherapy can secure control where re-excision would cost function.

- Q20 · A** Preoperative RT is 50 Gy in 25 fractions with surgery 4–6 weeks later. Postoperative doses are 60 Gy for a negative margin, 66 Gy for a positive margin and up to 75 Gy for gross residual disease. Shorter schedules remain investigational.
- Q21 · E** In NCIC SR2 local control and survival were equal, wound complications were 35% versus 17%, and late fibrosis was lower with preoperative RT, which is why it is preferred for most patients.
- Q22 · E** Retroperitoneal sarcoma (10–15% of soft tissue sarcomas) produces fullness, early satiety or leg symptoms late. Nodal spread is uncommon, and complete resection at the first operation is the only cure.
- Q23 · E** MAP is standard up to age 40: about 10 weeks before surgery, resumed a week after surgery to complete about 29 weeks.
- Q24 · B** Adding ifosfamide and etoposide to VDC improved 5-year event-free survival in localised disease (INT-0091). Adults receive the same protocol at full intensity.
- Q25 · A** Response is judged on the resection specimen, not on how much the mass shrinks.
- Q26 · B** Doxorubicin remains the reference first line. Combinations raise response rates but not clearly survival (EORTC 62012).
- Q27 · A** Doxorubicin causes cumulative cardiomyopathy, so echo or MUGA beforehand and tracking of the lifetime dose are needed, with precautions against extravasation.
- Q28 · B** Ifosfamide also causes encephalopathy and a proximal tubulopathy (Fanconi syndrome), so kidney function, phosphate and bicarbonate are monitored. Leucovorin is the rescue for methotrexate.
- Q29 · D** Pazopanib (PALETTE: PFS 4.6 versus 1.6 months, no overall survival gain) is for non-adipocytic sarcomas. Eribulin is the later-line drug with a survival benefit in liposarcoma.
- Q30 · D** Weekly paclitaxel is the first choice (ANGIOTAX). Checkpoint inhibitors are particularly active in cutaneous head and neck angiosarcoma.
- Q31 · B** Examination and chest CT are most frequent in the first 2–3 years, to catch lung metastases and local relapse while they can be resected. Retroperitoneal liposarcoma is the exception and recurs late.
- Q32 · C** Fertility preservation (sperm banking, or ovarian protection) must be arranged before starting treatment, and it is often forgotten in the rush to begin.
- Q33 · C** The plain X-ray is still the first and most useful bone test. MRI of the whole bone follows, and the patient should be referred before any biopsy.
- Q34 · E** Lymph nodes are rarely involved, so routine node dissection or sentinel biopsy is not used. In the subtypes named, examine the regional nodal basin; nodal spread then makes a limb or trunk sarcoma stage IV.
- Q35 · B** The scar and drain site are contaminated and must be treated; an unplanned excision enlarges both the radiotherapy field and the definitive operation. For stage III UPS, pembrolizumab with RT can be discussed (SU2C-SARCO32).
- Q36 · B** It is the first immunotherapy result in localised sarcoma. Offer it after shared decision-making where access allows; survival data are immature and it was not tested with chemotherapy.
- Q37 · E** Surgery alone is reasonable for superficial, ≤ 5 cm, low-grade tumours with clear margins (Yang 1998). Radiotherapy adds local control, but not survival, for most tumours that are over 5 cm or deep, and grade 2–3.
- Q38 · B** An adequate margin is not a single number. About 1 cm of fat or muscle, or an intact fascial plane, is usually enough with radiotherapy.
- Q39 · D** Image-guided preoperative RT to these reduced volumes gave few marginal failures and late grade 2 or worse toxicity of about 10%, showing that smaller margins are safe.
- Q40 · D** Myxoid liposarcoma spreads to soft tissue, the retroperitoneum and bone marrow without lung disease, so the spine and abdomen are imaged. It is exceptionally radiosensitive and trabectedin is active.
- Q41 · D** It supports dose de-escalation in an exceptionally radiosensitive tumour, but 50 Gy remains the standard outside trials.
- Q42 · E** Fat with a solid non-fatty nodule indicates dedifferentiation. Percutaneous posterior core biopsy of a retroperitoneal mass is safe and does not raise local recurrence; open or laparoscopic biopsy is avoided.

- Q43 · B** Postoperative RT is avoided because bowel falls into the tumour bed. Surgery by an experienced team remains the key treatment.
- Q44 · A** Extended resection improves local control in liposarcoma (Bonvalot 2009). Never enucleate, morcellate or “debulk” at the first operation, and refer before laparotomy.
- Q45 · E** Leiomyosarcoma metastasises to lung and liver, so both are scanned. Power morcellation of a fibroid with suspicious features should be avoided because it can spread an unsuspected sarcoma.
- Q46 · D** A bruise that will not fade on an irradiated breast is angiosarcoma until a punch biopsy says otherwise; reassess any “bruise” persisting beyond 2 weeks. MYC amplification supports radiation-associated angiosarcoma.
- Q47 · C** The disease is skin-led and multifocal. Re-irradiation (often hyperfractionated) and taxanes are used in selected cases.
- Q48 · A** The absolute risk is very low and the benefit of radiotherapy in breast cancer is large. Persistent skin changes should still be biopsied.
- Q49 · B** A poorly placed biopsy track can turn limb salvage into amputation, so plan the biopsy with the treating team. The whole bone is imaged for skip lesions, and baseline audiometry, echo and kidney function precede chemotherapy.
- Q50 · B** Fever, raised ESR and a bone lesion can be Ewing sarcoma. Send tissue for histology and culture together, and never curette a suspected sarcoma as if it were infection.
- Q51 · E** MAP is standard up to age 40. Where methotrexate cannot be monitored safely, carboplatin–ifosfamide–doxorubicin is an alternative, with more second leukaemias.
- Q52 · C** Adding ifosfamide–etoposide added toxicity only. Zoledronate (OS2006) and interferon maintenance for good responders (EURAMOS-1) were also negative.
- Q53 · B** If radiotherapy is the local treatment, chemotherapy continues during it with doxorubicin omitted. From age 18, cycles may be given every 14 or 21 days.
- Q54 · C** Interval compression with growth-factor support is the benefit. Higher doses per cycle (Granowetter 2009) and adding vincristine–topotecan–cyclophosphamide (AEWS1031) did not help.
- Q55 · B** Definitive RT is used when the tumour is unresectable or resection would be mutilating (spine, much of the pelvis, skull base). Postoperative RT is given for positive margins, gross residual disease or high-risk chest wall tumours.
- Q56 · B** PFS was 12 versus 6 months and overall survival 33 versus 24 months, but grade 3 or worse toxicity was 97%, so growth-factor support and LFT and CK monitoring are needed. Doxorubicin plus dacarbazine is an alternative.
- Q57 · A** Use the combination when shrinkage is the goal (symptoms, borderline resectable disease); otherwise single-agent doxorubicin is reasonable.
- Q58 · E** Doxorubicin alone is reasonable when palliation is the aim. Pazopanib is not used for adipocytic sarcomas.
- Q59 · A** PEComa shows TSC1/2 loss and chemotherapy has little activity. Watch for hyperglycaemia, stomatitis and pneumonitis.
- Q60 · D** TRK inhibitors give high, durable responses in any tumour type. Screen with pan-TRK IHC and confirm with RNA NGS; NTRK fusions are rare in adult sarcoma but common in infantile fibrosarcoma.
- Q61 · D** Selection explains much of the benefit: long disease-free interval, few nodules, controlled primary and no extrathoracic disease. Five-year survival after resection is about 30–40% in series.
- Q62 · A** Hydration, urine alkalinisation, drug levels and leucovorin rescue are routine. NSAIDs, PPIs and co-trimoxazole delay clearance, and glucarpidase is used for delayed clearance.
- Q63 · E** Audiometry and magnesium monitoring are needed with cisplatin, and kidney tubular damage follows ifosfamide and cisplatin. Anthracyclines cause cardiomyopathy and alkylators cause infertility and second cancers.
- Q64 · D** CT abdomen–pelvis and chest is done every 3–6 months for 2–3 years, then every 6–12 months. Leiomyosarcoma, by contrast, metastasises early.
- Q65 · D** MRI is preferred to CT for the primary site (better sensitivity, no radiation). Chest CT surveillance for lung metastases continues alongside.
- Q66 · C** Many young survivors walk, run and play sport. Late effects also include kidney damage, radiotherapy fibrosis, fracture and growth disturbance.

- Q67 · D** GIST is driven by KIT or PDGFRA (DOG1 positive); desmoid has nuclear β -catenin with CTNNB1 or APC changes and is observed first.
- Q68 · D** Preoperative and postoperative timing give similar survival, but preoperative treatment makes limb-sparing surgery easier and shows the histological response.
- Q69 · A** Chemotherapy before and after surgery raised long-term survival of localised osteosarcoma from under 20% to about 65–70%, making chemotherapy standard.
- Q70 · D** This established VDC/IE as the backbone of Ewing sarcoma chemotherapy.
- Q71 · A** This made VDC/IE the single international standard.
- Q72 · E** The meta-analysis suggested an 11% absolute survival gain, but the largest trial was negative, partly because it included small, low-grade and non-limb tumours. Benefit is modest and uncertain.
- Q73 · B** Three cycles were also as effective as five in the earlier trial (Gronchi 2016), so three cycles is the regimen used when chemotherapy is given.
- Q74 · B** Olaratumab is a lesson in confirming phase 2 signals with a phase 3 trial (Tap 2016).
- Q75 · D** Responses were seen mainly in those four types (SARCO28, Alliance A091401), irrespective of PD-L1.
- Q76 · D** Chemotherapy is ineffective. Pembrolizumab plus axitinib is used if the disease is bulky or symptomatic (Wilky 2019); pazopanib or sunitinib are later options.
- Q77 · B** Epithelioid sarcoma affects young adults, mimics a granuloma or ulcer, and spreads to lymph nodes; anthracycline first if unresectable.
- Q78 · A** EWSR1::ATF1 defines clear cell sarcoma, which is not a melanoma despite the immunophenotype.
- Q79 · A** Resect the primary and observe limited metastases; VEGFR tyrosine kinase inhibitors (sunitinib, pazopanib) are used if it progresses.
- Q80 · C** Surveillance comes first; systemic therapy is reserved for progression or symptoms.
- Q81 · C** ALK fusions occur in about half of cases, and resection is the first choice when feasible.
- Q82 · B** This is a desmoplastic small round cell tumour of adolescents and young men.
- Q83 · D** Small round cell sarcomas need molecular confirmation. Ewing sarcoma carries EWSR1::FLI1 (about 85%) or EWSR1::ERG (about 10%); CIC::DUX4 and BCOR::CCNB3 sarcomas are separate, more aggressive entities.
- Q84 · C** Le Deley 2010 found that fusion type does not change prognosis, but molecular testing is still needed to confirm the diagnosis.
- Q85 · A** SPEARHEAD-1 studied engineered T cells in synovial sarcoma and myxoid/round cell liposarcoma. Cytokine release syndrome and cytopenias are managed in a cell-therapy centre.
- Q86 · D** MDM2 and CDK4 amplification mark well-differentiated and dedifferentiated liposarcoma. Pazopanib is not used in adipocytic sarcoma.
- Q87 · A** This is why “pazopanib is not for liposarcoma; eribulin is.”
- Q88 · A** IDH1/2 mutations are found in most conventional central chondrosarcomas, but there is no established IDH-directed therapy outside trials.
- Q89 · E** Conventional chondrosarcoma is chemoresistant, but these two variants are treated by analogy with their chemosensitive counterparts.
- Q90 · C** Sarculator (Callegaro 2016) is used for extremity sarcoma; a histology-specific nomogram exists for the retroperitoneum (Gronchi 2013).
- Q91 · B** The biopsy track should still be placed where it will be removed with the tumour.
- Q92 · C** Standard preoperative RT remains 50 Gy in 25 fractions outside trials.
- Q93 · C** This is why radiotherapy is used for most large, deep, high-grade tumours, to protect the limb rather than to extend life.
- Q94 · A** Mifamurtide with MAP was studied in INT-0133 and is used where licensed.
- Q95 · D** Trials are preferred. Osteosarcoma TKI evidence comes from REGOBONE and CABONE.
- Q96 · E** High-dose busulfan–melphalan is used in some protocols for metastatic disease.

Q97 · C Periosteal osteosarcoma is intermediate and chemotherapy is debated. High-grade tumours, including those of the jaw, usually get cisplatin-based chemotherapy.

Q98 · C 35–40% of childhood rhabdomyosarcomas arise in the head and neck, and paediatric-type sarcomas always receive protocol chemotherapy.

Q99 · C MYC amplification supports radiation-associated angiosarcoma over a benign post-radiation atypical vascular lesion.

Q100 · C ALT above 3 times the upper limit of normal means interrupting the drug.

Q101 · E Trabectedin causes transaminitis, neutropenia and rhabdomyolysis; a rising CK or bilirubin means delaying or reducing the dose.

Q102 · B This subtype spreads to extrapulmonary soft tissue, retroperitoneum and bone marrow.