THE COLLEGES OF MEDICINE OF SOUTH AFRICA

Incorporated Association not for gain
Reg No 1955/000003/08

Examination for the Subspeciality Certificate in Medical Oncology of the College of Paediatricians of South Africa

21 February 2019

Paper 1 (3 hours)

All questions are to be answered. Each question to be answered in a separate book (or books if more than one is required for the one answer)

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1 An 18-month-old female patient presents to the paediatric surgeons with a 1-month history of constipation. The paediatric surgeon notices a swelling just above her buttocks and is worried about a possible mass. She has no peripheral nodes; no palpable abdominal masses and her BP is normal. She goes on to have a MRI of the lumbar spine, sacrum and pelvis which reveals a large mixed cystic and solid mass within the presacral region.

a) What is your differential diagnosis? (7)
b) What further investigations would you ask for? (8)
c) A biopsy of the mass reveals a mixed germ cell tumour with yolk sac elements. Her alpha feto-protein (αFP) is 50918 and her β-HCG is 1. The paediatric surgeon refers her to you. Describe how you would manage this patient. (13)
d) Write notes on tumour markers in germ cell tumours. (12)
[40]

2 A 14-year-old boy is referred from a peripheral hospital with a 4-week history of abdominal distention and vomiting. On examination his weight and height are within normal parameters, he has abdominal distention with ascites and a large central abdominal mass which measures 10 cm x 10 cm. In addition, he has significant cervical and supraclavicular lymphadenopathy. Blood tests reveal a WBC 13 x 10^9/L and platelet count of 53 x 10^9/L. His urea and creatinine are moderately elevated, LDH > 2000 IU/L and uric acid is 0.78 mM/L. and the HIV-Elisa test is negative. A biopsy of the cervical lymph node reveals a malignant process suggestive of a non-Hodgkin's lymphoma.

a) Briefly describe the staging of paediatric NHL. (10)
b) Discuss the immunophenotypic tests you would ask your pathologist to perform to delineate the type of NHL i.e. Burkitt lymphoma, diffuse large B-cell lymphoma, anaplastic large cell lymphomas and lymphoblastic lymphoma. (10)
c) Describe the cytogenetic findings in paediatric Burkitt lymphoma, diffuse large B-cell lymphoma, anaplastic large cell lymphomas and lymphoblastic lymphomas. (10)
d) Write notes on the pathogenesis of HIV-related lymphomas. (10)
[40]

3 You are asked to consult on an 8-year-old boy initially admitted with a swelling of his right mid-thigh and an abnormal x-ray. On clinical examination you find that he is pale. He has a single left supraclavicular lymph node and a painful swelling of the right thigh. His blood pressure is 142/93.

You review the investigations done to date:

Hb 7g/dL, MCV 81fl WBC 10.4 X 10^9/L Plts 324 X 10^9/L, ESR 124 mm/hr, ALT 12 IU/L, LDH 2014 IU/L, Ferritin 1011 mcg/L, urate 0.19 mM/L.

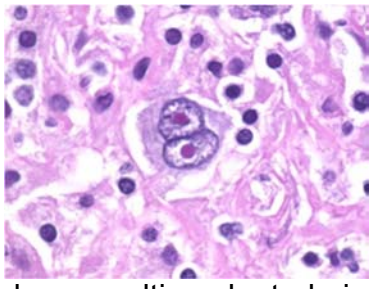
You review his chest x-ray sent with him from the referring day hospital. On first impression it looks normal but on secondary survey you notice a mass behind the heart on AP. You also notice that the fourth rib on the right and seventh rib on the left look moth eaten. Lateral chest x-ray confirms a posterior mediastinal mass (T7-T10). In addition, an ultrasound of the abdomen shows a second left paraspinal mass encasing adjacent vessels. Plain film x-ray of the femur shows an expanded mid-portion with a moth-eaten appearance to the bone.

a) What is the likely diagnosis at this point? Justify your answer. (6)
b) Describe your approach to his blood pressure control. (4)
c) Describe your approach to making the diagnosis and relevant staging investigations you would employ. (15)
d) Discuss cis-retinoic acid with respect to:
   i) Role in this disease. (2)
   ii) List 3 side effects. (3)

After a careful discussion with the parents regarding the extent of the disease and the prognosis, a decision is made to palliate the patient from the outset. Three months into palliation, he begins to deteriorate and his requirement for analgesic support escalates. His parents ask you about the possibility of adding medical cannabis to his pain regimen.

e) Write short notes on medical cannabinoids. (10)
[40]

4 A 12-year-old boy, previously well, presented to a general practitioner (GP) with a one-month history of fever and weight loss. He received two courses of antibiotics over the next 3 months, but he did not improve. By the time the GP refers him to you for a suspected malignancy, the boy had also developed enlarged neck glands and night sweats. There is no known TB contact. On examination the boy looks wasted. He is pale with cervical, supraclavicular and axillary lymphadenopathy of 3-4cm diameter. A liver is palpable 4cm below the costal margin and his spleen 3cm below the costal margin. A lymph node biopsy is performed, and the histology



shows multi-nucleated giant cells in a background of inflammatory cells. The giant cells have large bilobed nuclei and two large nucleoli. On immunohistochemistry CD15, 20 and 30 were positive in the giant cells.

a) What is the most likely diagnosis? (2)

PTO/Page 3/Question 4b)…

b) Discuss the special investigations that you would request and motivate why. (18)
c) List five auto-immune phenomena that are associated with this disease. (5)
d) The parents have read up about chemotherapy and are concerned about potential fertility. What would you advise? (7)
e) Six months after successfully completing his treatment, the boy presents again with lymphadenopathy and relapsed disease is confirmed. What treatment options would you discuss with him and his parents now? (8)

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Paper 2 (3 hours)

All questions are to be answered. Each question to be answered in a separate book (or books if more than one is required for the one answer)

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1 Briefly discuss anti-GD2 immunotherapy for neuroblastoma. [10]

2 Write short notes on Juvenile Xanthogranuloma. [10]

3 Discuss the long-term follow-up programme of a patient who received treatment for stage IV nephroblastoma (high risk/unfavourable histology). The patient did not receive any radiotherapy. [10]

4 Write notes on the diagnostic criteria for haemophagocytic lymphohistiocytosis. [10]

5 Write short notes on PRETEXT staging of hepatoblastoma using diagrams to illustrate your answer. [10]

6 Write short notes on infantile fibrosarcoma. [10]

7 Discuss the chemotherapeutic agent asparaginase with regards to preparations, mechanism of action and side effects. [10]

8 A 12-year-old girl with T-ALL on maintenance presents with unilateral hip pain, unable to weight bear. Write short notes on the diagnosis and treatment of avascular necrosis. [10]

9 A 4-year-old boy presents with a limb-threatening, biopsy proven Kaposiform haemangioendothelioma (KHE). You decide to treat the child with sirolimus (Rapamycin^{TM}). Discuss the indications, mechanism of action, monitoring and possible side effects of sirolimus. [10]

10 A 3-year-old girl who presented with single-system, bone only LCH relapses 7 months after her first line therapy is completed. She presents on this occasion with a rash, bicytopaenia and organomegaly. Discuss your approach to the patient and therapeutic options in relapse. [10]