A

Part 1 examination

MOCK PAPER- A Haematology First paper
Candidates must answer ALL questions
Time allowed: Three hours

H&T

1. A 2 Year old boy has severe haemophilia A and has been on prophylactic factor replacement since 7 months age. He weighs 15 Kg and is currently taking Advate 500 units 3 times a week. Recently despite good compliance he is getting repeated breakthrough bleeding. You have checked his 1hr post FVIII infusion levels and it is 0.04iu/ml. You suspect patient may have developed inhibitor.
    a. How would you investigate to confirm inhibitor to FVIII in this patient? Define Bethesda unit. 8
    b. Discuss treatment options available with the parents for managing the child's haemophilia and inhibitors. Explain risk and benefits of each option 10
    c. Your local haemophilia patient association has invited you to prepare a leaflet about recent advances in treatment of haemophilia A and B. Prepare a leaflet with recent advances in treatment (including treatments on horizon) their risks and pitfalls 7

TRANSFUSION

2. i. A 33 year old mother of two, is admitted on your ward receiving treatment for acute myeloid leukaemia. She has developed febrile neutropenia and is receiving broad spectrum IV antibiotics and antifungal medication. Recently her platelet count has remained below 20 and despite daily platelet transfusions the increment in platelet count is poor.
   Define platelet refractoriness, list likely causes and how will you manage the patient? 12

2. ii. You are contacted by the surgeons, on call. A patient on their ward has suffered from a severe reaction following a blood transfusion with hypotension, fever 39.5C, chills and generalised erythema. He suspects there may have been mislabelling of the sample as there is another patient with a similar name in the same bay. How would you investigate and manage this patient. Outline the steps you will put in place to ensure it doesn't happen again? 13

GEN HAEMATOLOGY

3. The obstetric team has contacted you for advice and management of a 26year old primi-gravida who is 19 weeks pregnant. Her recent FBC is Hb 112g/L, WCC 10.2X109/L, Platelet count 32 X 10e9/L. Blood film confirmed thrombocytopenia and normal morphology. Coagulation screen is normal. Previous FBC 4 years ago was normal. She has no h/o bruising or bleeding and her pregnancy is progressing well.
    a. List common causes of thrombocytopenia in pregnancy 8
    b. What are the likely possibilities in this lady and how will you investigate and manage? 7
    c. Outline her management plan for delivery, epidural/spinal anaesthesia, neonatal management and provide advice about thromboprophylaxis if hospitalised post-delivery. 10

MALIGNANT

4. A 70 year old female presents with haemoglobin 79g/L, WCC 15.6X10e9/L, Plat 29X10e9/L, peripheral film confirms myeloblasts and BM confirms tri-lineage dysplasia on morphologic appearance. Molecular genetic results on bone marrow aspirate confirm NPM wild type and Flt3 mutation. Negative for PML-RARA, CBFB-MYH11 and RUNX1-RUNX1t1.
   Pt suffers from Type 2 Diabetes and HT both of which are well controlled. Her performance status is 0.

    a. Briefly describe common cytogenetic and molecular genetic techniques used in diagnosis of AML. List common cytogenetic, molecular genetic abnormalities which impact treatment and prognosis. 8
    b. Outline the management plan for the patient with explanation of your decision making. 10
    c. Pt receives 4 courses of intensive chemotherapy and is in remission. She chose not to have a stem cell transplant. 10 months after stopping treatment she has relapse of AML. Discuss various treatment options and prognosis. 7