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6/6/2016

POSTGRADUATE INSTITUTE OF MEDICINE
UNIVERSITY OF COLOMBO

POSTGRADUATE DIPLOMA IN CLINICAL HAEMATOLOGY
EXAMINATION – JUNE 2016

Date: 6th June 2016 Time: 1.00 p.m. – 4.00 p.m.

Answer **four** questions only.
Answer each question in a separate book.
All questions carry equal marks.

PAPER I - ESSAY

1. A 78-year-old female with aplastic anaemia was admitted for red cell transfusions. Her Hb was 6.2g/dl.
   Two units were planned to be given on the same day. One hour after starting the second unit, she developed difficulty in breathing, tachypnoea and cyanosis. Discuss the possible causes for the above clinical manifestations and explain how you would differentiate them. (100 marks)

2. Discuss the pathophysiology, clinical presentation and management of Thrombotic Microangiopathy (TMA) syndrome. (100 marks)

3. A 50-year-old man diagnosed with chronic myelogenous leukaemia (CML) was managed with tyrosine kinase inhibitors (TKI). After six months he developed disease progression.

   3.1 Discuss the molecular basis of CML. (50 marks)

   3.2 What mechanisms are responsible for disease progression? (50 marks)

4.

   4.1 Discuss the pathophysiology of E/ß thalassaemia including the genetic factors which influence the severity of the disease. (60 marks)

   4.2 Outline the investigations that are done at the initial diagnosis and follow up of a symptomatic E/ß thalassaemia explaining the significance. (40 marks)

5. Discuss the pathophysiology of primary antiphospholipid syndrome (APLS) and critically evaluate the investigations used in the diagnosis of APLS. (100 marks)

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POSTGRADUATE DIPLOMA IN CLINICAL HAEMATOLOGY
EXAMINATION – JUNE 2016

Date: 7th June 2016 **Time:** 9.00 a.m. – 12.00 noon

Answer **six** questions only.
Answer each question in a separate book.
All questions carry equal marks.

PAPER II
STRUCTURED ESSAY QUESTIONS (SEQ)

1.

1.1 List the important haematological parameters which can be obtained from modern haematology analyzers. (20 marks)

1.2 Briefly discuss the need to examine a blood film prior to releasing a full blood count report, despite the availability of fully automated analyzers. (35 marks)

1.3 Briefly describe the methods available for internal quality control (IQC) and external quality control (EQC) for the full blood count. (25 marks)

1.4 Explain the interpretation of Levey-Jennings charts. (20 marks)

2. A diagnosed patient with acute leukaemia with a low platelet count received several platelet transfusions without an adequate increment of the platelet count. A diagnosis of platelet refractoriness was suspected.

2.1 How do you assess the response to platelet transfusions in this patient to establish the diagnosis? (30 marks)

2.2 What are the different causes for platelet refractoriness? (20 marks)

2.3 Outline the management of suspected immunological platelet refractoriness. (35 marks)

2.4 Give three measures that can be adopted to prevent this complication? (15 marks)

Contd..../2-

3.

3.1 List the causes of megaloblastic anaemia in children. (15 marks)

3.2 Describe the biochemical basis of megaloblastic anaemia. (30 marks)

3.3 Describe the investigations you would do in a child with clinical features of megaloblastic anaemia. (35 marks)

3.4 Outline the management of megaloblastic anaemia in children. (20 marks)

4.

4.1 Describe the classification of Myelodysplatic syndrome (MDS). (40 marks)

4.2 What are the cytogenetic abnormalities in myelodysplastic syndrome (MDS). (10 marks)

4.2 Describe the risk stratification for prognosis of MDS. (25 marks)

4.3 Outline the treatment options available for MDS. (25 marks)

5. A previously well 55-year-old man presented with fatigue, fever and cough of two weeks duration.
The full blood count showed white blood cell count of 50,000/cmm^{3}with 20% blast cells, normal haemoglobin level and normal platelet count. The C-reactive protein (CRP) level was 400 mg/L (Normal <6mg/L). Bone morrow cytogenetics showed the following karyotype: 46XY, inv (16) (p13.q22). Acute myeloid leukaemia (AML) was diagnosed and treatment commenced.

5.1 Discuss the prognostic relevance of the above given data on the treatment outcome of this patient. (15 marks)

5.2 Describe the molecular basis of the karyotypic defect found in this patient and describe the morphological changes in the peripheral blood and bone marrow. (15 marks)

5.3 Discuss the importance of cytogentic factors in AML prognosis. (40 marks)

5.4 Write a brief account on **two** mutations described in AML. (30 marks)

Contd..../3-

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6. A 60-year-old man was investigated for pallor and loss of weight.
His full blood count is as follows:
   - Haemoglobin level – 9g/dL
   - WBC Count – 4.0 x10^9/L (Neutrophils – 2.0x10^9/L, Lymphocytes – 1.6x10^9/L, Monocytes – 0.4x10^9/L)
   - Platelet count - 160x10^9/L

   Blood picture – Marked rouleaux formation seen. Red cells – normochromic normocytic. White cells and platelets – appear normal.
   Biochemistry investigations:
   - Creatinine level – 2.4 mg/dL (Normal 0.6 -1.2 mg/dL)
   - Serum protein electrophoresis – Comment: Monoclonal band (region IgG).
   - M-protein quantification – (IgG) 15g/L

   6.1 What further investigations you would do to arrive at a diagnosis of a plasma cell disorder? (25 marks)

   6.2 Briefly describe the prognostic markers of multiple myeloma. (30 marks)

   6.3 Discuss the role of adhesion molecules in multiple myeloma. (20 marks)

   6.4 Describe the pathogenesis of renal failure in multiple myeloma. (25 marks)

7. A 55-year-old female was found to have the following full blood count results:
   Hb level – 13 g/dL
   White cell count – 9x10^9/L (Neutrophils – 6.0x10^9/L, Lymphocytes – 2x10^9/L, Eosinophils – 1.0x10^9/L)
   Platelet count – 900 x10^9/L

   7.1 List the investigations you would do to arrive at a diagnosis. (20 marks)

   7.2 How would you differentiate essential thrombocythaemia from other types of myeloproliferative neoplasms based on the peripheral blood and bone marrow morphology. (40 marks)

   7.3 How would you risk stratify essential thrombocythaemia (ET)? (20 marks)

   7.4 Diagnosis was confirmed as ET. Outline the management of this patient. (20 marks)