Examination for MD Degree
Course Title: Commentary
Date: 25/11/ 2020
Time allowed: 90 minutes

TANTA UNIVERSITY
Faculty of Medicine
Pediatric Department

Total assessment: 200 Marks

A 14-year-old boy was referred to the pediatric clinic for short stature and excessive weight gain. History revealed that weight gain started from the age of 4 years and slow growth was noticed especially over the last year. He was born at 39 gestational weeks with a birth weight of 2750 g. His parents were not related. His father was diagnosed with hypertension at the age of 32 years and died due to intracranial hemorrhage at 37 years of age. His paternal grandfather was hypertensive.

At presentation, his weight, height, and body mass index were 55.1 kg, 143.3 cm ,and 26.8 kg/m2, respectively. Target height was 167.6 cm. He had acanthosis nigricans and did not have any striae or buffalo hump. Cardiac and abdominal examinations were unremarkable. His pubic hair was Tanner stage 3 and testicular volumes were 6 ml. He had mild developmental delay and speech disturbance. Blood pressure (BP) was 150/90 mmHg. Fasting plasma glucose (4.7 mmol/L), serum potassium (6.4 mmol/L), serum sodium (138 mmol/L), serum creatinine (42.4 µmol / L), and fasting lipid profile were normal at initial evaluation. 24-h free urinary cortisol and overnight dexamethasone test were normal. spironolactone treatment (100 mg/12h) was initially commenced. but the patient's BP remained high for age during this treatment.

Further investigations reveled serum potassium, 7.2 mmol/L; chloride, 113 mmol/L; pH, 7.28 - HCO3, 17.7 mmol/L), serum creatinine, (46.8 µmol / L). Fundoscopic examination, renal doppler ultrasonography, and echocardiography were all normal. Aldosterone and renin concentrations before treatment gave the following results: 90 pg/ml. and 0.01 pg/ml, respectively. Hypertensive load by ambulatory blood pressure monitoring was >50%

Comment

Examination for MD Degree
Course Title: pediatrics (paperII)
Date: 18/11/ 2020
Time allowed: 3hours
All Questions should be tried

TANTA UNIVERSITY
Faculty of Medicine
Pediatric Department

Total assessment: 260 Marks

Long Assay

Q1: Ciliopathies **(50 marks)**

Short Assay

Q2: Common agents potentially toxic to young children in small doses (25 marks)

Q3: Intestinal transplantation in children with intestinal failure (25 marks)

Q4: Anemia of chronic disease (25 marks)

Q5: Drug-induced abnormalities in Folate metabolism (25 marks)

Q6: Refeeding syndrome (25 marks)

Q7: Discuss briefly conditions associated with disorders of

gluconeogenesis in infants and children (25 marks)

Q8: MCQs, *(3 marks for each question):*

1. A child appears with a chronic fatigue, recurrent epistaxis, bone pain, and bruise that occurs easily. A physical examination reveals a pale child with hepatosplenomegaly and generalized tenderness over the long bones. The ENT findings are normal. The laboratory studies reveal anemia, thrombocytopenia, and elevated liver function tests. The long bone x-ray reveals an Erlenmeyer flask deformity of the distal femur. The most likely diagnosis is:

a) GM1 gangliosidosis
b) Fabry disease
c) Niemann-Pick disease type 1
d) Gaucher disease type 1

2-Which of the following is the correct recommendation for a patient receiving antihypertensive treatment with alpha adrenergic blockers for pheochromocytoma?

a) 2 L fluid restriction
b) High salt diet
c) Low salt diet
d)Low salt and low potassium diet

3- A previously healthy 8-month-old girl with 1st attack of RSV infection. Which of the following mechanisms will most likely operate to clear the infection?

a) CD4+ T cell-mediated necrosis of infected cells
b_Cytotoxic T cell-induced apoptosis of infected cells)
c) MHC I presentation of viral peptides on Cos+ T cells
d) Virus-specific antibodies that neutralize free virus

4-A 3-month-old male infant has recurrent infections and is found to have an impaired ability to kill microbes by the nitroblue tetrazolium test. Which of the following conditions is most likely responsible for the findings in this patient?

a) Chediak-Higashi syndrome
b) Chronic granulomatous disease
c) Hereditary angioedema
d) HIV/AIDS

5- A 6-year-old male receives a bone marrow transplant from his father during treatment for acute myelogenous leukemia. Of primary concern will be the potential development of:

a) acute rejection.
b) an allergic reaction.
c) autoimmune responses.
d) graft-versus-host disease.

6- 13-yr-old child presented with fever and altered sensorium with MRI showing medial temporal lobe involvement suggests:

a) Neisseria
b) HSV
c) Cerebral malaria
d) Fungal meningitis

7- Of post resuscitative efforts listed below, the one shown to have efficacy in the cardiac arrest patient who has demonstrated return of spontaneous circulation (ROSC) is administration of:

a) Glutamate antagonists.
b) Calcium channel antagonists.
c) Free radical scavengers.
d) Insulin to control hyperglycemia.

8-You are caring for a 17-year-old adolescent in septic shock. Despite aggressive crystalloid fluid resuscitation, he remains hypotensive. Your vasopressor of choice would be:

a) Dopamine.
b) Dobutamine.
c) Nordopamine.
d) Epinephrine.

9- You are managing a trauma adolescent with a flail chest who is in respiratory distress. Indications for mechanical ventilation would include:

a) Respiratory rate >25/min.
b) Respiratory rate <12/min.
c) Pao2 <70 mm Hg at Fio2 >0.5.
d) Paco2 >55 mm Hg at Fio2 >0.5.

10- A 2-year-old patient with biliary atresia underwent an orthotopic liver transplantation 1 month ago. Now, he presents with elevated bilirubin, alkaline phosphatase, and transaminase levels. Acute rejection versus biliary obstruction is considered. Which are the histologic features used to diagnose acute cellular rejection?

a) Mixed portal inflammation, portal/central vein endotheliitis, and bile duct injury
b) Mixed portal inflammation, cholestasis, and bile ductular proliferation
c) Lobular inflammation, cholestasis, and bile duct injury
d) Lobular inflammation, hepatocyte apoptosis, and increased plasma cells

11- A 10-day-old neonate presents with an elevated direct bilirubin level. Biliary atresia is excluded on cholangiogram. A liver biopsy is subsequently performed. The patient's hepatologist asks the pathologist if the neonate may have alpha-1antitrypsin deficiency. What would be the pathologist's response?

a) No globules are seen, so the patient does not have α1-antitrypsin deficiency.
b) No globules are seen, but I cannot exclude α1-antitrypsin deficiency in a neonate.
c) α1-Antitrypsin deficiency is not in the differential diagnosis of biliary atresia.
d)There are no characteristic pathologic findings in α1-antitrypsin deficiency.

12-A 25-year-old woman presents with acute fatty liver of pregnancy at 35 weeks of gestation. Following emergency caesarean section delivery of a male infant, she recovers rapidly. Which of the following investigations will be the MOST informative?

a) Maternal blood acylcarnitine analysis
b)Infant blood acylcarnitine analysis
c) Maternal liver biopsy
d)Maternal urinary organic acids

13-A 5-year-old with a history of hereditary spherocytosis presents with pallor and fatigue. One week ago, he was seen by his primary care doctor for a fever and runny nose. He was diagnosed with an upper respiratory tract infection at that time. His mother reports that his fevers and runny nose resolved, but over the past several days he has developed progressive fatigue and pallor. His vital signs are as follows: temperature 36.7, heart rate 160, RR 26, and BP 110/54. On examination, you note an extremely pale child. A complete blood count reveals:

- **White blood cell count: 13 × 10 9 /L**
- **Hemoglobin: 5.4 g/dL**
- **Hematocrit: 16.2%**
- **Platelets: 164 ×10 9 /L**
- **Reticulocyte count: <1%**

What test will likely reveal an underlying etiology?

a) Parvovirus B19 PCR

b) Direct antibody testing

c) Bone marrow aspiration

d) PTT

14- In examining a 4-year-old girl who is new to your practice, you discover that she has rudimentary thumbs and is well below the 5th percentile for both weight and height. You also observe irregular hyperpigmentation on the trunk and anogenital areas. Of the following, the MOST likely hematologic disorder associated with these findings is:

a) Bloom syndrome
b) Diamond-Blackfan anemia
c) Fanconi anemia
d) Thrombocytopenia and absent radii (TAR) syndrome

15-You are evaluating a 12 year girl who was admitted to the hospital for anemia (hemoglobin concentration of 85 gm/L) who has had significant vaginal bleeding with the onset of menarche 3 weeks ago. Her family history includes several females who were diagnosed with von Willebrand disease (VWD). Which of the following tests are necessary to detect the presence of type 2A VWD?

a) Von Willebrand factor (VWF) antigen concentration
b) Ristocetin cofactor activity
c) Factor VIII activity
d) Multimer analysis (VWF)

16- Regarding the defect in late ribosomal biogenesis, all of the following is true except

a) Causes Pancytopenia, mainly neutropenia
b) Exocrine pancreatic insufficiency
c) Causes pure red cell aplasia
d) Hypoplastic bone marrow

17-Leprechaunism includes all of the following features except:

a) Fasting hypoglycemia
b) Postprandial hyperglycemia
c) Severe resistance to insulin
d) Large for gestational age

18- All of following critical values obtained at a time of fasting hypoglycemia (plasma glucose less than 50 mg/dL) suggest hyperinsulinism except:

a) Plasma insulin level is more than 2 μU/mL
b) Plasma free fatty acids are more than 1.5 mmol/L
c) Plasma beta-hydroxybutyrate level is less than 2.0 mmol/L
d) Plasma insulin level is 4 μU/mL

19-The preferred screening test for patients with a chronic granulomatous Disease is:

a) G6PD qualitative assay
b) RBC G6PD activity
c) Dihydrorhodamine 123 fluorescence (DHR) test
d) Ability of neutrophils to kill microorganisms

20. All of the following are manifestations of septo-optic dysplasia except:

a) Nystagmus
b) Optic nerve dysplasia
c) Large anterior pituitary gland
d) Incomplete development of septum pellucidum

----------Good Luck----------

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