TEACHING FILES

Grand Rounds

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EXTRAHEPATIC PORTAL HYPERTENSION

Clinical Problem : A 12 year old body born of non consanguineous marriage presented with hematemesis. Oesophageogastroscopy {OGDscopy} showed portal hypertension. Liver functions were normal. Colour Doppler of abdomen showed multiple collaterals. Child was diagnosed as a case of extrahepatic portal hypertension and underwent splenic artery embolization as he had recurrent hematemesis inspite of Sclerotherapy. He continued to have hematemesis with signs of hypersplenism and thus was operated for splenectomy with mesocaval shunt. After the surgery, child again had 2 episodes of hematemesis and varices on OGDscopy.

Question : Why did the child have hematemesis inspite of shunt surgery?

Expert Opinion : Hematemesis is portal hypertension occurs due to bleeding from varices. Varices are porto-systemic anastomosis that occur to relieve the portal pressure. If the portal pressure is very high, then bleeding can occur as the varices burst. Sclerotherapy is a palliative therapy in which varices that look impending for bursting are sclerosed to prevent bleeding. Splenectomy with shunt surgery help to decrease portal pressures as well as create an artificial shunt between systemic and portal circulation to relieve the portal pressures. Thus, in a child with shunt surgery and normal liver, the prognosis is good. However in a child with liver disease, there is always a risk of precipitating hepatic encephalopathy as the toxic metabolites tend to bypass the liver and do not get degraded and can affect the brain. A repeat bleeding post shunt surgery denotes that the portal pressures have again increased. This suggests occlusion of the shunt and the child should be investigated for the same. In this child, colour doppler was done and it was found that mesocaval shunt was not visualized suggesting that shunt was blocked. The child is again on regular sclerotherapy.

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SIX MONTH WITH PALLOR

Clinical Problem : A 6 month old boy born of non consanguineous marriage presented with increasing pallor for 2 months and abdominal lump noticed 5 days back. There was no fever. The child is 3rd of three siblings and other two children are normal. Birth, immunization and milestones are normal. He is on breast feeds plus weaning diet. On examination, he had tachycardia, pallor, hepatosplenomegaly and cardiomegaly. Other systems were normal. Investigations showed:

- Hemoglobin = 3.4 gm, dl {MV = 67.6, MCH = 19.7, MCHC = 29.1}
- WBC count = 100,900 cumm {15 percent neutrophils, 83 percent lymphocytes}
- ESR = 70 mm at end of 1 hour
- Liver enzymes = Normal
- Lipemic serum with LDH = 4746 IU L and uric acid = 12.8 mg percent
- Peripheral smear = nucleated RBCs.
- HIV = Negative

Question : What is the diagnosis?

Expert Opinion : This child has anemia with high WBC count. The peripheral smear shows nucleated RBCs which may be counted as WBC on the coulter machine which may cause false elevation of the WBC count. Nucleated RBCs are immature red cells which are precursors of the mature RBCs. {In RBC maturation process in the bone marrow, the earlier erythrocytes have a nucleus which is lost as the cell matures into a reticulocyte and a mature RBC}. Thus, in peripheral blood, nucleated RBCs are usually not seen. Presence of nucleated RBCs is suggestive of increased destruction {hemolysis} of RBCs which makes the bone marrow throw immature cells into the circulation. This hemolysis can be in the spleen or the bone marrow. The lipemic serum with high LDH and uric acid is again suggestive of a high cell turnover and may be seen in leukemia or hemolysis. However, in leukemia, nucleated RBCs are not seen. Thus, in this child one would suspect hemolytic anemia. The child's corrected WBC count was 15,900 cumm after excluding the nucleated RBCs and hemoglobin electrophoresis showed presence of beta thalassemia in the child {HbF = 70 percent, HbA2 = 1.8 percent} with both father {HbA2 = 6.3 percent} and mother HbA2 = 5.9 percent} being thalassemia carriers.

Thus, a WBC count of over 1,00,000 cumm is not always suggestive of leukemia or leukemoid reaction and examination of the peripheral smear is always a must to look for abnormality in morphology of RBC and WBC.

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HOLI COLOUR INHALATION

Clinical Problem: A 10 years old boy presented with accidental inhalation and ingestion of golden metallic coloured powder during Holi festival followed by difficulty in breathing within 10 minutes of the episode associated with giddiness. He vomited once and the vomitus contained golden yellow substance. On examination, he had mild to moderate respiratory distress, yellow staining of face, bilateral fine crepts, drowsiness and respiratory rate of 42/min. Other systems were normal. Investigations showed:

- Hemoglobin = 13.5 gm/dl
- WBC count = 16,700/cumm (83% polymorphs, 13% lymphocytes, 3% eosinophils)
- Platelet count = 3,23,000/cumm
- Bilirubin = 2.3 mg% (Direct = 0.5%)
- SGOT = 42 IU/L, SGPT = 26 IU/L
- USG Abdomen = Normal.

His bilirubin normalized after 3 days and prothrombin time and Partial thromboplastin time was normal. A repeat CBC after 6 days was normal. Chest X-Ray