TEACHING FILE

Grand Rounds

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BUDD CHIARI SYNDROME

Clinical Problem: : A 3 months old girl presented with ascites. There is no jaundice. Ultrasound abdomen with colour doppler showed ascites with hepatomegaly with non-visualisation of hepatic veins. CT angiography showed thrombosis of all 3 hepatic veins suggestive of Budd-Chiari syndrome (BCS).

How should such a child be managed?

Expert Opinion : The traditional approach to treatment involves systemic thrombolysis in all BCS patients without a specific contraindication. Surgical portosystemic shunt or TIPS is performed in progressive cases of BCS or as a bridge to transplantation. Patients with acute or fulminant BCS are best served with liver transplantation. Recently, there have been an increasing number of successful reports involving endovascular techniques including angioplasty and stent placement specially for hepatic webs. (1) Surgical shunting can lead to chronic hepatic encephalopathy. Balloon angioplasty of HV thrombosis has shown variable results throughout the world, restricting its use to short-segment HV thrombosis only. Angioplasty in infants is difficult due to small size of vessels. Thus, the choice of therapy would be difficult in this child considering the age. This child underwent dilatation of hepatic veins transjugularly but had no response. She then underwent portocaval shunt but continued to have ascites. A liver biopsy at time of surgery showed nodular liver with cirrhosis. She was subsequently advised liver transplant but could not undergo same and died by 10 months of age.

References

1. Bozorgmanesh A,Selvam D,Caridi J. Budd-Chiari Syndrome: Hepatic Venous Web Outflow Obstruction Treated by Percutaneous Placement of Hepatic Vein Stent. Semin Intervent Radiol. 2007; 24: 100–105

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Quick Response Code

NEONATAL CHOLESTASIS

Clinical Problem : : An 8 months old boy born of non consanguineous marriage presented with jaundice, yellow urine and clay stools since day 6 of life. On examination, he had rickets, weight was 6.2 kg, He had jaundice with hepatosplenomegaly. His liver biopsy was suggestive of biliary atresia.

How to take care of his nutrition and deficiency of fat soluble vitamms?

Expert Opinion : Since Vitamins A, D, E and K are fat soluble vitamins and require bile for absorption, in case of biliary atresia, due to cholestasis, bile is not secreted into the intestines and thus there is malabsorption of fat soluble vitamins. Thus, these vitamin deficiencies can occur. Supplementation of these vitamins in 10 times recommended daily allowance is thus needed. Water soluble forms of these vitamins should be administered orally so that absorption in absence of bile is not hampered. Regarding nutrition, since bile excretion is poor, even fats are not absorbed. Only medium chain triglycerides (MCT) are absorbed. Thus supplements with MCT oil is required. Adequate calorie and protein intake should be taken care of so that nutrition requirements are met.

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FEVER WITH RASH

Clinical Problem: A1½ year old girl presented with fever for 15 days and erythematous rash 12 days ago that lasted for 5 days. There is no cough, vomiting, loose motions or altered sensonum. She was treated with multiple antibiotics but fever did not respond. On examination, vital parameters were normal. She had red lips with peeling of skin over the hands. Other systems were normal. There was no lymphadenopathy.

What is the diagnosis?

Expert Opinion : This child had fever with rash. Fever with rash may be due to infectious causes, autoimmune disorders, vasculitis, drug reaction or even malignancy. In most viral exanthemas usually the fever subsides with onset of rash or along with the rash. It is very unusual for the fever to persist even when rash has disappeared. Rash with malignancy is usually due to petechiae or purpura and has associated other bleeding tendencies, anemia and bone tenderness with organomegaly. Drug reaction can also lead to rash but rash is usually itchy and subsides with antihistamines. It may be urticarial. Drug hypersensitivity can lead to Steven Johnson syndrome in which case mucocutaneous wide spread eruptions with desquamation will be present which is not there in this case. Thus autoimmune disorder or vasculitis is the most likely cause. Autoimmune disorders with fever and rash as presentation may be systemic onset JIA. However there would be subsequent joint involvement, organomegaly or lymphadenopathy which is not seen in this child.

Thus one should suspect vasculitis. Common vasculitis in children is Kawasaki disease associated with fever, rash, strawberry tongue, cervical lymphadenopathy, conjunctival congestion and subsequent desquamation of skin. In this child, all features are present except cervical lymphadenopathy and conjunctival congestion. However, it is not necessary that all features of Kawasaki disease be present as some may evolve over time. Thus, most likely diagnosis in this child is Kawasaki disease. In this child, echocardiography showed coronary dilatation, ESR was 120 mm at end of 1 hour and platelet count was 8,00,000/cumm. Treatment with intravenous immunoglobulin (IVIG) led to resolution of fever.

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