TEACHING FILE

GRAND ROUNDS

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TUBERCULOUS MENINGITIS WITH OCCLUSION OF MIDDLE CEREBRAL ARTERY

Case:- A 7 month old girl was diagnosed as tuberculous meningitis (TBM) with hydrocephalus in April 2011 in view of fever for 15 days and convulsions. She was started on 4 drug antituberculous therapy (ATT) and steroids. Her CSF at that time showed 180 cells/cumm (90 percent polymorphs, 10 percent lymphocytes) with proteins of 142 gm/dl and sugar of 23 mg%. Her CT brain showed basal exudates with hydrocephalus and ischemic infarcts in lentiform, caudate nucleus and anterior temporal lobe. Chest X-Ray showed right upper lobe consolidation. Her mother was also on ATT for past 2 months. The child had spasticity, dystonia and had lost all previous milestones. She was on doing well till Feb 2012 following which she developed increasing dystonia. A repeat CT scan showed large gliotic area in left basal ganglia and left frontotemporal periventricular white matter with moderate hydrocephalus and right middle cerebral artery (MCA) was not visualized but left MCA was totally occluded. She was advised to continue same. ATT and aspirin was added.

Why did the child get a vascular occlusion?

Expert's opinion: Vascular changes consisting of arterial narrowing with or without occlusion are frequently seen at autopsy in cases of tuberculous meningitis. (1) These changes lead to necrosis of localized areas of the brain and may be responsible for focal neurological deficits. (2) Vessel involvement only at the base of the brain, without peripheral vessel involvement, is strongly suggestive of tuberculous meningitis as this is the area that bears the brunt of inflammation in tuberculous meningitis. (2) The walls of the vessels immersed in thick gelatinous exudate are affected by inflammation and arteritis leading to vascular occlusion and thrombosis. Thus vascular occlusions are not uncommon in tuberculous meningitis. .

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EXTRAHEPATIC PORTAL VEIN OBSTRUCTION WITH HOMOCYSTINURIA

Case: - A 2½ year old boy presented with abdominal distension for 2 years. This was preceded by episode of vomiting that required hospitalization and intravenous fluids for 8-10 days. There was no hematemesis, malena or jaundice. On examination, weight was 11.6 kg. He had splenohepatomegaly. Other systems were normal. Ultrasound (USG) of abdomen with colour doppler of portal system showed prominent caudate lobe of liver with coarse echotexture, splenomegaly, non-visualization of portal vein and numerous collaterals at splenic hilum, around gall bladder and along anterior abdominal wall. Splenic vein and superior mesenteric veins were also not seen. Liver function tests showed bilirubin of 0.5 mg/dl, SGOT – 47 IU/L, SGPT – 37 IU/L, albumin of 2.8 gm/dl. Esophageogastroscopy (OGDscopy) showed single grade 2 varix. Liver biopsy was normal. His antiphospholid antibody, anticardiolipin antibodies were negative and Protein C, Protein S, antithrombin III levels were normal. Urine for homocysteine was positive. Ophthalmological evaluation showed no lens dislocation, echocardiography was normal, serum vitamin B12 levels were normal (417). He was continued on propranolol and folic acid.

Is this homocystinuria?

Expert's opinion: Homocystinuria is an amino acid disorder. Most individuals are lacking, or have low function, of the enzyme cystathionine beta-synthase (CBS), leading to a buildup of methionine and homocysteine in the body. Clinical manifestations include growth delays, psychiatric disturbances, delayed developmental milestones, and learning disabilities or intellectual disabilities. If untreated, homocystinuria can cause lens dislocation and glaucoma, osteoporosis, scoliosis, heart disease or stroke due to thrombi, or pancreatitis. Mutations in the CBS, MTR (methionine synthase), MTRR (methionine synthase reductase), and MTHFR genes cause homocystinuria. Presence of homocystinuria in urine may be false positive. Thus a positive test must be confirmed with serum methionine levels. A positive screening test depends upon protein ingestion. False positive screens is present (these include heat-damaged specimen, or antibiotic therapy). Methionine levels of up to 2 µ mol/mL characterize cystathionine synthase deficiency. In our patient, serum methionine was not elevated, thus, it is unlikely to be homocystinuria. His repeat urine for homocystinuria was negative after 1 month of folic acid.

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SPUTUM POSITIVE MILIARY TB IN A CHILD WITH DOWN'S SYNDROME

Case: - A 13 years old boy with Down's syndrome presented with intermittent evening rise of temperature for past 2 months associated with productive cough. He had lost 4 kg weight in the same time and had loss of appetite. He had received a 9 month antituberculous

treatment (ATT) at 7 months of age and again a one month ATT at 10 years of age for pericardial effusion. There was no contact with tuberculosis patient. He was immunized till date. He had delayed milestones and was currently studying in 1st grade and could take care of himself. On examination, his weight was 36 kg, height was 135.5 cm. He had pallor, clubbing, mongloid features and insignificant cervical lymphadenopathy with hypotonia and hepatomegaly. Other systems were normal. Investigations showed hemoglobin of 9.2 gm/dl, white cell count of 13,000/cumm (78 percent polymorphs, 19 percent lymphocytes), ESR of 105 mm at end of 1 hour. Mantoux test was negative and sputum for acid fast bacillus (AFB) was positive for AFB bacillus. Chest X-Ray showed miliary mottling. Patient was started on category II ATT regime and TB culture sensitivity after 6 weeks showed sensitivity to all antituberculous drugs. After 3 months of ATT, sputum for AFB was negative and ATT was stopped after 1 year of therapy. Child had gained 10 kg in one of treatment.

How common is presence of acid fast bacilli in the sputum smear of a miliary TB?

Expert's opinion: In adults with suspected tuberculosis, the sputum smear is a helpful test. For a positive acid-fast bacilli sputum test, at least 5,000 to 10,000 bacilli/millimeter of specimen are required. (1) Mycobacterium tuberculosis culture needs 10-100 microorganisms. (2) Patients with miliary tuberculosis usually do not have cavities. Therefore, they present a very low concentration of bacilli when compared to those with cavitatory TB. (3) Thus, though chances of getting a bacteriological confirmation in miliary TB is high with a culture, presence of acid fast bacilli on a sputum smear in children with miliary TB is rare. In immunocompromised patients, the bacterial load is high and chances of getting a positive AFB on smear in miliary TB is high. (4) Our patient had Down's syndrome which leads to poor immune response and thus he seems to have had a positive AFB.

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