TEACHING FILE

GRAND ROUNDS

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RECURRENT ENCEPHALOPATHY

Case Report: A 6 years old girl born of non consanguineous marriage presented with projectile vomiting since 1 day followed by irrelevant speech, rowdy behavior and uprolling of eyes and clenching of teeth 8-10 times. There was no history of fever. She had a similar episode 1½ years back and was diagnosed as acute Cerebellitis. At that time CT brain and CSF examination were done which were normal. She also had an episode of vomiting followed by limpness a month back. In between these episodes she is alright though her scholastic performance has decreased since past 1½ years. She achieved all her milestones properly and birth history is normal. Family history is non contributory. On examination, she had bradycardia with irregular respiration suggestive of raised intracranial tension that improved with Mannitol. She was conscious but disoriented. Speech was slurred and there were no cranial or focal neurological deficits. Pupils were bilaterally equal and reacting to light. She had hypertonia with brisk reflexes and extensor planters. Other systemic examination was normal.

What is the cause of her recurrent encephalopathy?

Expert's opinion :-

Lactic acidosis. This child has presented with an acute episode of encephalopathy. Common causes of encephalopathy include infections {encephalitis}, substance or drug abuse, lead poisoning, metabolic disorders, demyelination and space occupying lesions. Of these, substance abuse, demyelinating disorders and metabolic disorders present with recurrent episodes. Encephalitis usually may be acute or lead to an insidious chronic presentation. It will not present with recurrent episodes with interspersed normal episodes in between. Similarly space occupying lesions may present with seizures or signs of raised intracranial tension and rarely cause no deterioration over a period of time.

This child has presented with recurrent episodes of disorientation, vomiting and limpness. Her pupils are normal. In case of substance abuse, pupils are one of the clinical markers that give a suspicion of drug abuse. The pupillary reflex is maintained in case of metabolic causes of coma. Atropine causes fixed, dilated pupils; morphine results in small pupils which react normally to light and barbiturates give rise to fixed pupils. Meiosis is seen with organophosphates, opiates and phenothiazines whereas mydriasis is seen with antihistamines, tricyclic antidepressants, alcohol and cocaine. Also with no other evidence of autonomic involvement, substance abuse seems unlikely. Lead poisoning presents with encephalopathy which is not an intermittent variety.

Thus, one needs to consider demyelinating disorders or metabolic disorders in this child. Demyelinating disorders usually cause focal neurological deficits as they predominantly involve white matters. This child has altered behavior which is suggestive of grey matter involvement in addition to hypertonia and brisk reflexes suggestive of white matter involvement. Thus, the most likely cause of encephalopathy in this child is a metabolic cause.

This child's blood glucose was normal. Her urine ketones were negative. She had metabolic acidosis with increased anion gap. Her serum ammonia was marginally elevated [178 µg/dl (Normal: 30 to 90 µg/dl)] and her lactate was 33 mg/dl with pyruvate of 0.32 mg/dl and lactate: pyruvate ratio of 103. MRI brain showed hyperintense signals in cerebral white matter and dorsal aspect of pons suggestive of mitochondrial or organic acidemia. Her urine for toxic screen was negative and muscle biopsy showed irregular muscle fibres. Her urine organic acids and aminoacidogram was normal. Thus she was diagnosed as Lactic Acidosis with recurrent encephalopathy.

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POLIOMYELITIS

Case Report: A one and half year old unimmunized boy developed sudden onset lower limb weakness 2 months ago. On examination, he had hypotonia, weakness more in extensors of the calves, bilateral foot drop and absent reflexes. Other systems were normal. Stool grew polio virus on culture.

Should this child be given vaccination for polio vaccine?

Expert's opinion: -

This child has acute flaccid paralysis due to polio. There are 3 types of polio viruses. Since this child is unimmunized, he is susceptible to all 3 types of polio viruses. Since, he has a polio infection at the moment; he is going to develop natural immunity against one virus type. However, he is still susceptible to other 2 types of polio virus. Thus, the child should be given polio vaccine to develop immunity against the other 2 types of polio viruses. He should receive polio vaccine after 4 weeks of resolution of the acute illness.

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ANEMIA, RICKETS AND RECURRENT DIARRHEA

Case Report: - A 2½ years old boy presented with abdominal distension and recurrent diarrhea for 6 months. Stools were frothy and had oil droplets. He also had a blood transfusion 2 month ago. At that time he had black coloured stools. There was no fever, bleeding from any site. Bone marrow examination at that time was normal. Doctor noticed bow legs at that time and gave 6 lakhs of Vitamin D. There was no jaundice, exposure to chronic drug abuse or altered sensorium. Diet was balanced mixed diet and immunization and milestones were normal. On examination, he was found to have palmar erythema, double malleoli, bow legs and hepatosplenomegaly with dilated tortuous abdominal veins with flow away from umbilicus. Other systems were normal.

With is the cause of his symptoms?

Expert's opinion:

This child has organomegaly with palmar erythema suggestive of liver disease. He has dilated tortuous abdominal veins with flow away from umbilicus and splenomegaly suggestive of portal hypertension. Malena could be due to bleeding from varices. Malabsorption in form of frothy and oil droplets in stools suggests fat malabsorption which could be due to liver or pancreatic disease. Fat malabsorption leads to poor absorption of fat soluble vitamins such as Vitamin A, D, E & K. This could explain the rickets in the child. Anemia needing blood transfusion could be due to bleeding from varices or hypersplenism. Thus, the cause of problems in this child is chronic liver disease with portal hypertension even though child has never had jaundice

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