TEACHING FILE

GRAND ROUNDS

Ira Shah

A 14 MONTHS OLD BOY WITH SUDDEN ONSET PARAPLEGIA

Case:- A 14 months old boy born of third degree consanguineous marriage presents with sudden inability to stand or sit since 15 days. There is no restriction of movement in upper limb. There is no convulsion, bladder or bowel complaints and no trauma. The child has fever since 3 days. His milestones and immunization history is adequate. There is no history of TB contact. On examination, the child has hypertonia in both lower limbs with decreased power and brisk reflexes in both knees. There is bilateral sustained ankle clonus and planter reflex was extensor. Cremasteric and abdominal reflexes were absent. Upper limb reflexes were present. Pain and touch sensation appears intact. Other systemic examination is normal.

What is the diagnosis?

Expert's opinion:- This child has presented with sudden onset paraplegia. One may consider possibilities such as poliomyelitis, trauma, Guillian Barre Syndrome (GBS), transverse myelitis and spinal tumor which may have bled suddenly leading to intraspinal edema. Since the paralysis is symmetrical and the child has been immunized upto date, poliomyelitis seems unlikely. Trauma and vertebral facture is also unlikely as no history of trauma is available. Since the paralysis has remained static, GBS also seems unlikely as GBS is usually an ascending polyneuropathy. A sudden onset paraplegia can be a manifestation of transverse myelitis but it usually leads to a band of dermatomal involvement with band like pain at the level of the lesion and complete motor, sensory as well as bladder and bowel involvement below the upper level of involvement. In this child, bladder and bowel involvement cannot be commented upon as the child has still not achieved continence. However, pain and touch sensation are intact thus making a diagnosis of transverse myelitis less likely.

Spinal tumors usually have an insidious onset of paraparesis. However, they may be totally asymptomatic and can present as sudden onset paraplegia if there is bleeding inside the tumor. Thus, one would consider a possibility of spinal tumor in this child.

An MRI spine in this child was suggestive of right paravertebral soft tissue mass at the level of T1 to T5 spinal vertebrae extending into the spinal canal through T1 & T2 neural foramen with marked extradural compression of spinal cord suggestive of intraspinal tumor.

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ASCITIS

Case: - A 12 years old boy presented with progressive abdominal distension since 8 months and edema feet for 1 week. There is no jaundice, bleeding from any site or fever. He was treated with antituberculous therapy (ATT) for 3 months but had no relief. There is no history of blood transfusion or any other disorder in past. On examination, he has dilated veins over abdomen with flow below-upwards, massive ascitis and hepatosplenomegaly.

What is the diagnosis?

Expert's opinion: This child has ascitis along with dilated veins. The normal flow of blood over abdomen is away from umbilicus (i.e., below upwards above umbilicus and above downwards below umbilicus). In postal hypertension also, the flow is away from umbilicus but veins are dilated and tortuous. In inferior vena cava (IVC) obstruction the flow of blood is from below upwards (i.e., below upwards both above and below the umbilicus). In this child too, flow of blood is below-upwards suggestive of IVC obstruction. With development of ascitis, the diagnosis is Budd Chiari syndrome. The color Doppler of abdomen confirmed obstruction in the inferior vena cava.

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DROOLING OF SALIVA

Case: - An 11 years old give presented with unsteadiness while walking for 1 year, change in voice and dribbling of saliva since 6 months. There is no neuroregression.

What is the diagnosis?

Expert's opinion: This child has unsteadiness while walking and change in voice suggestive of either a cerebellar problem or a basal ganglia problem. Since there is drooling of saliva, it is suggestive of basal ganglia involvement. Now basal ganglia diseases that come with unsteadiness while walking and change in voice are Parkinson's disease, Wilson's disease, rheumatic chorea and Huntington's chorea. Both rheumatic chorea and Huntington's chorea would present with choreoathetoid movements which this child does not have. Parkinson's disease would present as mask like facies, gait disturbances and slurred slow speech, however dribbling of saliva is not usually present. Neurological manifestations of Wilson's disease usually have drooling of saliva, and above mentioned symptoms. Thus most likely diagnosis in this child is Wilson's disease. On further evaluation, she had Kayser-Fleischer (KF) rings and liver dysfunction. Her 24 hours urine copper was 159 mg in 24 hours which increased to 1033mg after penicillamine challenge test. Thus, she was diagnosed as Wilson's disease.

From: Medical Sciences Department, Pediatric Oncall, Mumbai.

Address for Correspondence Dr Ira Shah, 1/B Saguna, 271, B St Francis Road, Vile Parle (W), Mumbai 400056.

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