

Case Report

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First manifestation of severe haemophilia: acute flaccid paralysis due to spinal cord contusion with subdural hemorrhage

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ABSTRACT

Hemophilia A and hemophilia B are the most common and serious congenital coagulation factor deficiencies. Intracranial hemorrhages occur in 3-10%. Spinal epidural hematomas are rare. Even with severe hemophilia, only 90% have evidence of increased bleeding by 1 year of age. Only 2% of neonates with hemophilia sustain intracranial hemorrhages. Here we describe a case of a 5-month old boy with positive family history of hemophilia A on his maternal side who was admitted to the hospital because of retention of urine and decreased movement in both lower limbs following history of fall from height (approximately 2 feet high), two days prior to admission. Physical examination showed no skin lesion or hematoma. Neurological examination showed the infant in frog like posture with flaccid paralysis of both lower limbs without loss of sensation. There was associated bladder involvement with a Phantom hernia on the right side of his abdomen. MRI Spine revealed cord contusion in D4-D9 segment with spinal subdural hemorrhage. Coagulation profile was abnormal with a prolonged Activated plasma thromboplastin time. Factor VIII assay revealed a level of 1%. Treatment was conservative, and the infant was given factor VIII replacement. There was remarkable improvement within weeks. Thus, spinal hematomas being rare should still be considered and ruled out for prompt management of cases of suspected hemophilia. This case highlights the importance of a thorough family history which led to the ultimate diagnosis of severe Hemophilia A by coagulation profile and neuroimaging and further confirmed by factor VIII assay.

Keywords: Acute flaccid paralysis, Cord contusion, Subdural hemorrhage

INTRODUCTION

Deficiencies of factors VIII (Hemophilia A) and IX (Hemophilia B) are the most common severe inherited bleeding disorders. They are X linked Recessive disorder. Neither factor VIII nor factor IX crosses the placenta; bleeding symptoms may be present from birth or may occur in the fetus. Only 2% of neonates with haemophilia sustain intracranial hemorrhage, and 30% of male infants with haemophilia bleed with circumcision. Even in patients with severe haemophilia, only 90% have evidence of increased bleeding by 1 year of age.¹ Central

nervous system bleeding in Haemophilia A is quite frequent whereas intraspinal bleeding is very unusual.^{2,3} The laboratory screening test that is affected by a reduced level of factor VIII or factor IX is PTT. In severe haemophilia, the PTT value is usually 2-3 times the upper limit of normal. Results of the other screening tests of the haemostatic mechanism (platelet count, bleeding time, prothrombin time, and thrombin time) are normal.

CASE REPORT

A 5-month-old male child hailing from Bihar, born out of non-consanguineous marriage, was brought to our

Emergency room of upgraded Department of Pediatrics, PMCH with the chief complaint of dribbling of urine and decreased movement of lower limbs following fall from bed (approximately 2 feet high) two days prior to admission to our hospital. There was no history of fever, loose stool or recent immunization or any other recent illness. On thorough history taking it was discovered that there was a case of severe Haemophilia on the maternal side of the family.

On clinical examination, all vital parameters and anthropometry were normal. Developmental quotient in all four domains were within normal limit. The child is on exclusive breast feeding. The mother gave no history of easy bruising or hematomas following i.m. injections during routine immunization nor prolonged bleeding anytime. CNS examination revealed a conscious child with flaccid paralysis of both lower limbs. He maintained a frog like posture and his bladder was distended (Figure 1).



Figure 1: Infant in frog like posture due to the flaccid paralysis of the lower limb.



Figure 2: Phantom hernia on the right side of abdomen due to weakened abdominal wall muscles.

He also had a Phantom Hernia on the right side of his abdomen (Figure 2). All other systemic examination was normal. Plain X-ray of pelvis with both hip was then done and found normal. Ultrasound whole abdomen revealed an over distended bladder as patient was unable to void, otherwise a normal scan. The child was then catheterized. MRI Spine revealed cord contusion in D4-D9 segment with spinal subdural hemorrhage (Figure 3 and 4).

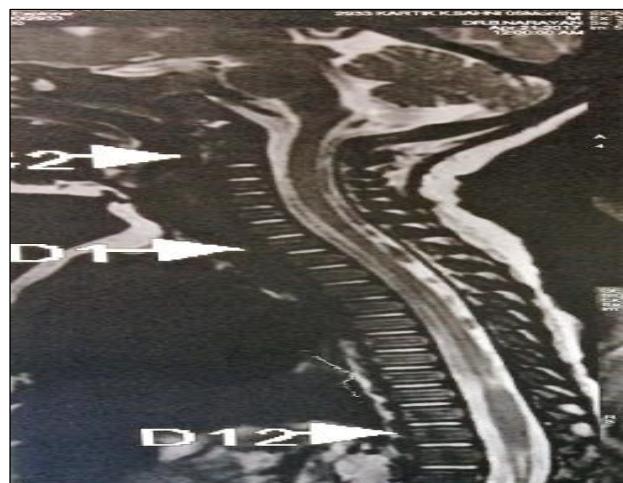


Figure 3: MRI cervicodorsal spine showing subtle linear streak hyperintensity (T2WI) involving the lateral part of upper and mid dorsal spinal cord (D4-D9 segment) suggestive of cord contusion.

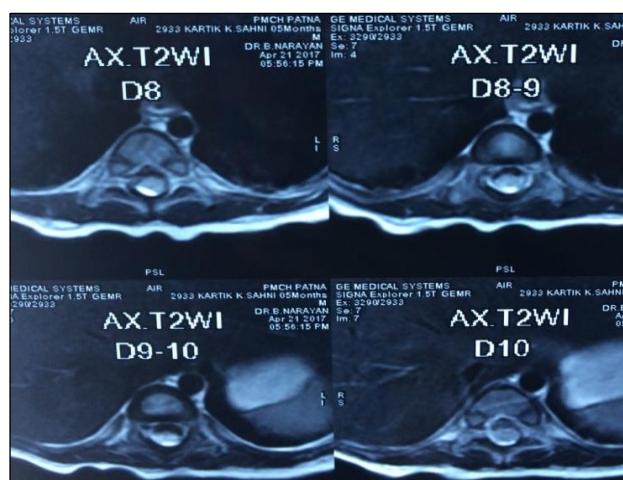


Figure 4: MRI transverse section of D8, D8-9, D9-10 and D10 showing linear band of T2 hyperintensity noted along the anterior and posterior subdural space of the spinal cord most likely spinal subdural hemorrhage.

The coagulation profile was discovered to be abnormal. Activated plasma thromboplastin time was prolonged i.e. 90.8 seconds (reference range 26-31 seconds) prothrombin time was normal i.e. 10.9 seconds (reference range 13-15 seconds). Factor VIII assay revealed a level of 1% only. Platelet count was 5.2 lacs/microlitre. Other

hematological parameters were normal. Serum electrolytes were normal.

The child was managed conservatively with infusions of factor VIII concentrate. There was remarkable improvement by the end of one week with gradual improvement in the strain of urine and increased in power of the lower limbs.

DISCUSSION

Spontaneous CNS bleed is an uncommon manifestation of hemophilia and is most frequent after the fourth or fifth decade. It is usually precipitated by trivial trauma. Intraspinal bleeds account for 2-8% of CNS hemorrhages.³ It is very rare but may present as the first manifestation of severe haemophilia in children.^{4,5} Signs and symptoms are often atypical in infants. In a literature review, 19 cases of pediatric hemophilic population were reported with spinal hematomas but none were from India.⁶ The most common site of a spontaneous spinal epidural hematoma is the cervicothoracic region or thoracolumbar region.⁷ The pathogenesis of spinal bleeding is believed to be secondary to rupture of epidural veins in the spinal epidural space. Intra-abdominal hemorrhage is often in the differential diagnosis of a spinal hematoma. In the present case, the 5-month-old male infant presented to us as a case of acute flaccid paralysis due to spinal cord contusion with subdural hemorrhage detected on MRI spine. Diagnosis of Hemophilia A was made after taking proper history, clinical examination and doing the coagulation profile and estimating the serum factor VIII level. Patient improved on conservative management and infusions of Factor VIII concentrate.

Recent reviews support treatment of selected cases with mild or stable neurological deficit conservatively with factor VIII concentrate infusions begun as early during the course as possible.^{3,8-10} Neurological recovery is gradual and complete recovery eventually occurs over weeks based on severity. Surgical evacuation of the hematoma by decompression laminectomy has its complications but maybe considered in case of delay at diagnosis or if conservative management fails or the condition worsens rapidly.^{2,3}

CONCLUSION

Thus it is important to always take a thorough family history and to have a high degree of suspicion and do early neuroimaging (CT or MRI) in every pediatric patient, especially male, presenting with signs of acute flaccid paralysis for accurate diagnosis and early intervention.

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