Spinal Epidural Hematoma in a Patient with Hemophilia B Presenting as Acute Abdomen

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Abstract Spontaneous spinal epidural hematoma (SSEH) is a rare complication in patients of haemophilia. We report the case of a 9-year-old boy with severe haemophilia B who presented with acute abdomen of 5 days duration. Acute onset of neck/back pain, walking impairment and urinary retention has usually been described as symptom complex in SSEH. The hematoma was identified by magnetic resonance imaging of the spinal column. Our case calls attention to recognition of abdominal pain (with no other localizing features) as initial symptom of SSEH and prompt evaluation and management before more overt symptoms of spinal cord compression becomes evident.

Keywords Hemophilia B · Epidural hematoma · Acute abdomen

Introduction

Spontaneous spinal epidural hematoma (SSEH) is a rare complication in hemophilia. The spontaneous development of spinal epidural hematoma is most frequent after the fourth or fifth decade [1]. However, it has been reported to occur in all age groups, and it is a very rare clinical entity in children. Only 30 pediatric cases of SSEH have been documented in the medical literature [2, 3]. Almost all of these cases have presented with neurological manifestations. We report a 9-year-old boy with hemophilia B who presented as acute abdomen and the cause for his symptom

on further evaluation was ascertained to be SSEH. To the best of our knowledge this association of acute abdominal pain with SSEH (very rare in hemophilia) and has not been reported in literature.

Case Report

A 9 yr old boy a known case of hemophilia B presented to us with abdominal pain in periumbilical region since 5 days. There was no history of trauma, diarrhea, fever, vomiting, skin or orificial bleed and joint swelling. There was no loss of consciousness, headache and seizures. A thorough physical examination found no abdominal guarding or rigidity or tenderness with no signs of any neurologic deficit. However while walking he used to assume a particular stooped posture. He had last received his factor IX concentrate 5 months back. Ultrasound abdomen and thighs was done at admission (to rule out psoas or retroperitoneal/intraabdominal bleed/pancreatitis) was normal. At admission his haemoglobin was 9.0 g/dL, total leukocyte count was $7.3 \times 10^9/L$, differential leukocyte count revealed polymorphs 60 %, lymphocytes 37 %, monocytes 2 % and eosinophils 2 % and platelet count was $376 \times 10^9/L$. A peripheral smear showed no basophilic stippling or sickle cells. Serum Amylase level was 29 (23–85 U/L). LFT, KFT, Calcium were within normal limits.

On second day of admission child developed difficulty in walking and urinary retention with paraspinal muscle spasm. Muscle power in lower limbs became 4/5. Deep tendon reflexes in lower limbs were hypoactive with positive babinski however there was no sensory loss. Magnetic Resonance Imaging of spine was planned in view of neurologic findings. The same evening he developed flaccid paralysis of both lower limbs.

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Fig. 1 MRI spine depicting SSEH from C4–T10 level

MRI Spine showed extensive spinal epidural hematoma extending from C6–T12 level of spinal cord [Fig. 1]. He was given Factor IX concentrate and urgent neurosurgical decompression was done.

Unfortunately the child succumbed to postoperative complications.

Discussion

CNS bleeding is the most dreaded life-threatening haemorrhage seen in people with haemophilia. Incidence of CNS bleeding in patients with haemophilia is variously quoted as 2.2–7.8 % [4]. Intraspinal bleed could be intramedullary, subdural or epidural.

The usual clinical presentation of a spontaneous spinal epidural hematoma is sudden stabbing neck or back pain that progresses toward paraparesis or quadriparesis, depending on the level of the lesion and the nerve root. Children often suffer from additional symptoms of irritability, and occasionally urinary retention. The male/female ratio of SSEH is 1.4 : 2 [4]. In high cervical region, SSEHs could cause spinal shock which is a fatal condition [5]. The association of SSEH

presenting as acute abdomen in children has not been reported earlier despite extensive search of literature.

Intraspinal bleeding in patients of hemophilia is rare. In a series of 2,500 patients with haemophilia over a period of 11 years, the prevalence rate of CNS bleeding for factor VIII and factor IX deficiency was 2.7 and 3.6 %, respectively. Out of 71 patients in this series who had CNS bleeding, 65 had intracranial while only 6 (8.4 %) had intraspinal bleeding [4]. All those with intraspinal bleed had initial presentation with neurological symptoms. None, even in this large series reported acute abdomen as initial presentation. The youngest hemophiliac to be affected is reported to be a 3 month baby who presented with increased irritability [6]. The other reported cases in literature presented with neurological signs, walking difficulty and neck/back pain [7–9]. Spinal epidural bleeding is presumed to be due to the rupture of the epidural veins. Lack of valves and minor resistance of these vessels can result in bleeding if a sudden increase in intra-abdominal or intrathoracic pressure occurs. Epidural hematoma has not been reported as a cause of abdominal pain in literature. Abdominal pain is a very unusual presentation of epidural hematoma in children with clotting disorders and is due to

entrapment of spinal nerves originating from vertebral foramina in the blood column.

The management of SSEH involves a judicious and aggressive use of factor replacement as well as surgical treatment in selected cases to optimize recovery [10, 11].

Patients of hemophilia should be screened for presence of intraspinal bleed when they present as a case of acute abdomen with no apparent cause for this symptom.

Conflict of interest None.

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