Case Report

Spontaneous Spinal Intramedullary Hematoma in an Infant: A Rare Entity and a Diagnostic Challenge

Vibhu Shankar Parashar, Priyanka Aswal¹, Tarun Kumar Gupta, Gaurav Jaiswal

Department of
Neurosurgery, RNT
Medical College, Udaipur,
¹Department of Pathology,
Geetanjali Medical College,
Udaipur, Rajasthan

ABSTRACT

Introduction: Spontaneous spinal intramedullary hematoma is a rare cause of acute paraplegia in adults and is extremely uncommon in children. Very few cases with no apparent etiology (such as trauma, vascular lesions) have been reported in adults. We did not find any apparent cause for the hematoma in our patient and to the best of our knowledge, this is first case reported in infants.

Case Report: We present the case of a 6-month-old female child admitted with acute-onset paraplegia, bladder bowel involvement, and no history of trauma or bleeding diathesis. The MRI showed an intramedullary mass extending from the D11-L1 level. The mass was excised and histopathology revealed it to be an organizing hematoma. Conclusion: Our case highlights that though it is a rare entity, there is a need for more awareness when dealing with children with sudden paraplegia, acute retention of urine, or neurological deficit. Early diagnosis and prompt surgery are crucial to achieve the best neurological outcome.

KEYWORDS: Idiopathic cause, intramedullary location, organizing hematoma, rare entity, spontaneous spinal hematoma

INTRODUCTION

Spontaneous intramedullary hemorrhage is a rare disease entity, and most cases have specific etiologies such as trauma, vascular malformations, and tumorous lesions or anticoagulant treatment.[1-4] It is extremely rare for this to occur without an apparent underlying etiology, and only a few such cases have been reported in adults earlier.[5] Surgical intervention should be performed as soon as possible to preserve and restore neurological function. Repeated hemorrhage can occur; close observation of patients after surgery is, therefore, important in cases without an apparent etiology. Recently, we came across a case of spontaneous intramedullary hematoma in a 6-month-old female child, which is, to the best of our knowledge, the first case to be reported in an infant without any attributable cause.

CASE HISTORY

We present a case of a 6-month-old female child who was admitted to our tertiary care center with acute-onset paraplegia with bladder bowel involvement. There was no history of trauma or any bleeding diathesis. Four days earlier, the child presented with episodes of crying with abdominal distension, inability to pass urine, and sudden-onset weakness in both lower limbs. On examination, the child was unable to sit without support but was able to lift her chest off the ground. Postural reflexes were absent.

Investigations

Hemogram and urinalysis was normal. Lumbar puncture revealed clear cerebrospinal fluid without any cells. An MRI of the spine showed an intradural, intramedullary well-defined mass lesion (25mm × 14 × 12mm) extending...
from D11-L1 level, which appears iso- to hypointense on T1W and hyperintense on T2W sequence [Figures 1 and 2]. On inspection of the spinal cord, no abnormal findings, such as abnormal vessels or discoloration, were found. Excision of the mass was performed, the mass was sent for histopathological examination, and histopathology confirmed it to be an organizing hemorrhage without any evidence of vascular malformation [Figure 3]. Therefore, intra-operative inspection and the pathology report did not find any pathology that might have caused hemorrhage. Postoperatively, she improved and regained bladder control.

**DISCUSSION**

Spontaneous intramedullary hematoma is a rare condition, and there have been only a few case reports seen in adults in the literature.[5-10] To the best of our knowledge, this is the first case reported in an infant. The most common etiology for this entity is spinal

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**Figure 1:** MRI of lumbosacral spine shows intramedullary lesion with T1W sequence without contrast

**Figure 2:** MRI of lumbosacral spine shows intramedullary lesion with T1W sequence with contrast
trauma, with others being vascular malformation, cavernoma, tumor, syrinx, bleeding disorders, and use of anticoagulants. In our patient, none of the etiologies just mentioned were attributed and labeled as idiopathic, that is, without any cause, which makes this case unique. Other atypical presentations such as mimicking a myocardial infarction or Brown-Sequard Syndrome have also been reported.[11,12]

Because spontaneous intramedullary hemorrhage with an unknown etiology is very rare, meticulous inspection of the hemorrhagic site to find the cause is essential. Repeated hemorrhage can occur[13]; therefore, close observation of patients after surgery is important in cases without an apparent etiology. An MRI is considered the most valuable diagnostic tool.

There are also a few other cases of spontaneous spinal hematomas with location in the epidural space described in literature. Spontaneous spinal epidural hematoma (SSEH) is an idiopathic accumulation of blood in the vertebral epidural space. It is a very uncommon cause of spinal cord compression. Cases without known predisposing factors are known to be spontaneous. SSEH is extremely rare in children and is mostly located in the dorsal epidural space. The cervicothoracic region is the most common location in such children. Ventral SSEH is even rarer, with only a few previous reports in children.[14-17]

Surgical removal of the hematoma is the treatment of choice, regardless of whether the course of hematoma is acute, subacute, or chronic. Immediate surgery may improve the prognostic outcome. The most important prognostic factor is the preoperative neurological status of the patient. Our patient was on follow-up and showed good recovery, although no recovery after the evacuation of hematoma has been reported in other studies.[10]

**Conclusion**

Our case highlights that since it is an extremely rare entity in children and infants and has varying clinical presentation, it remains a diagnostic challenge.[10] Meticulous inspection of the hemorrhagic site to find the cause is essential. Therefore, there is a need for more awareness when dealing with children with sudden paraplegia with acute retention of urine or any neurological deficit that may precede in spontaneous spinal hematomas.

Any delay in the diagnosis and treatment may cause permanent neurological deficit. Early accurate diagnosis and urgent surgical management of this rare entity may result in decreased morbidity and better neurological outcome.

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**Conflicts of interest**

There are no conflicts of interest.

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**Figure 3:** Photomicrograph showing microscopic findings of an organizing hematoma on H&E stain
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