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Case report

Spinal subdural hematoma revealing hemophilia A in a child: A case report

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Abstract

Background: Intraspinal bleeding especially in the form of subdural hematoma is rare in hemophiliacs. In the present case, we report a neglected hemophilic A child with such a problem and discuss its management options.

Case Presentation : A 9-year old hemophilic A boy presented with quadriparesis, confusion and meningismus after a fall 4 days previously. There was no sign of direct trauma to his back. His CT Scan and MRI showed spinal extramedullary hematoma extended from C5 to L2. We corrected the factor VIII level, but two days later, the patient's lower limbs weakened to 1/5 proximally as well as distally. We performed a laminectomy from T11 to L2, according to the level of the maximal neurological deficit and recent deterioration course. The subdural hematoma was evacuated. The hematoma in other spinal levels was managed conservatively. In the week following the operation, the patient's neurological status approached normal.

Conclusion: This case calls attention to the clinical manifestation, radiological features and management options of the rarely reported intraspinal hematoma in hemophilic children. Although this case has been managed operatively for its hematoma in the thoracolumbar region, at the same time it can be considered a successful case of conservative management of intraspinal hematoma in the cervicothoracic region. Both conservative and surgical management could be an option in managing these patients considering their neurological course.

Background

Hemophilia is an inherited hemorrhagic disease caused by a deficiency of a clotting factor. Hemophilia A is an X-linked recessive disorder caused by a deficient factor VIII and accounts for 85 percent of hemophilic cases.

Although central nervous system bleeding is a leading cause of morbidity and mortality among hemophiliacs, intraspinal bleeding especially in the form of subdural hematoma is extremely rare. In the present case we report a neglected hemophilic A child with quadriparesis due to such an intraspinal bleed.

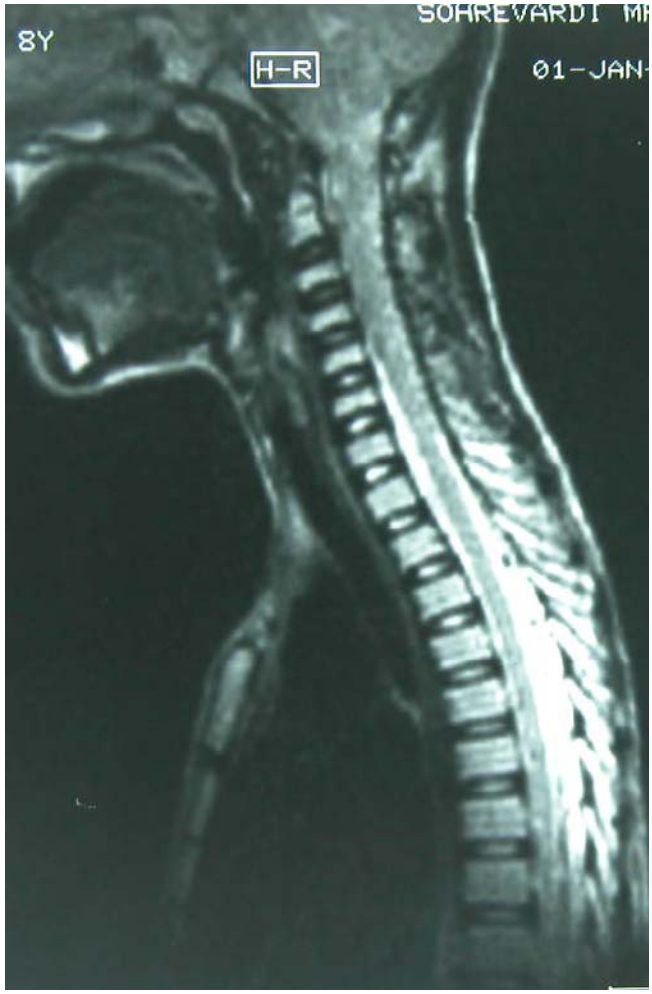

Figure 1
Saggital T2-weighted MRI of the cervicothoracic lesion. The extramedullary hematoma is extended from C5 to midthoracic.

Case Report

A 9-year old boy presented with quadriparesis, after a fall from a 2-meter height, 4 days prior to admission. He was confused and had nausea, neck and back pain and fever. Initially a reliable history was unobtainable. Subsequently, the family related that following any minor trauma, the patient would have joint swelling and ecchymosis for more than 5 days. He also had prolonged bleeding after dental extraction. He was confused (Glasgow Coma Scale = 13) and quadriparetic (upper limb 4/5, lower limb proximal 3/5 and distal 4/5) with hypoactive reflexes in four limbs and no sensory loss or urinary control problem. There was tenderness over diffuse levels of the spine, but mainly in his cervical and lower thoracic region. The patient had multiple ecchymotic lesions in his left upper limb but not on his back. Despite the quadriparesis, a CSF study through lumbar puncture was performed on the admission in view of the obvious neck rigidity, fever and lack of reliable history. The results were within normal limit. His emergency spine CT scan showed an acute extramedullary intradural hemorrhage in the spinal canal, from C5-L2 especially in the thoracolumbar region. In his MRI (Fig. 1,2), the extent of the hematoma could be clearly identified. No vascular malformation was seen. Further angiographic study was not performed. His brain imaging was within normal limit. Since the coagulation tests of the patient proved to be consistent with hemophilia A, we tried to correct the factor VIII level to normal (above 50%) and keep the patient under observation. 2 days later the patient deteriorated neurologically. Even though, his coagulation tests were within normal limits, his lower limbs weakened to 1/5 proximally as well as distally. His upper limb power did not change significantly at this time. The decision was made to operate on the patient. We extended the laminectomy to 4 levels from T11 to L2, according to the level of the maximal neurological deficit and recent deterioration. We realized then that we could have evacuated the hematoma through limited laminectomy. The dura was found to be tense and on opening, a significant amount of hematoma was evacuated through our rather small dural incision. The arachnoid and pia seemed to be intact. In spite of extension of the laminectomy, we tried to be minimally invasive. No abnormal bleeding was encountered during the operation. After hemostasis, the patient's dura was closed routinely. No fusion of the laminectomy levels was performed.

Postoperatively, the child did not improve for about three days. Afterwards, he began to improve gradually. Two days after operation, the patient had no fever. One week postoperatively, his lower limb force was within normal limit and his upper limb improved significantly. We tried to keep his factor VIII level above 30 percent for about 2 weeks postoperatively. 6 months later he was painless and his neurological examination was completely normal. His MRI showed complete resorption of hematoma in the whole spinal axis. The patient will be followed for possible spinal deformity in the future.

Discussion

Intraspinal bleeding in the hemophilic patients is rare. De Tezanos Pinto et al [1] reported only two cases of intraspinal bleeding among his patients (1410 hemophiliacs) between 1960 and 1991. In another 11-year study from 1965 to 1976, there were 6 patients with intraspinal bleeding in a population of 2500 hemophiliacs [2]. The incidence of intracerebral bleeding was 65 out of these 2500. Among these, spinal subdural hematoma is rare. We could find only 1 case of spinal subdural hematoma

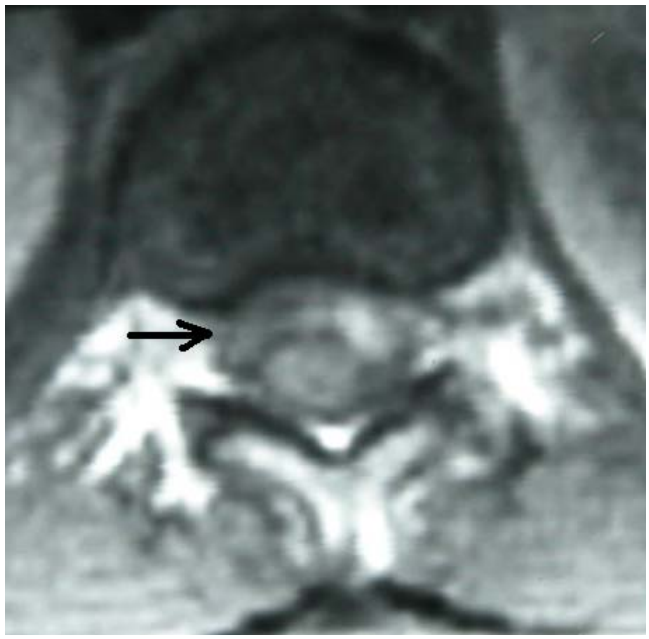

Figure 2
Axial Thoracolumbar MRI. The posterior fat pad and the hyperintensity rim around the anteriorly located hematoma (arrow), due to methemoglobin formation is compatible with subdural hematoma.

in the hemophilic patients reported on Medline [3]. The source of intraspinal bleeding in the hemophiliacs is rarely found. But, in patients who come to operation or autopsy, the majority of bleeds are found to be extramedullary, and only a small number are intramedullary [4]. 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. Spinal subdural hematomas cannot be explained through this pathogenesis. Vinters et al [5] have postulated that in many cases, spinal subdural hematomas may be due to a primary lesion that eventually dissects into the subdural space. As the subarachnoid blood is dissipated by CSF, the subdural clot is all that remains. Our patient's clear CSF study doesn't support this mechanism.

According to the works of Haines et al [6] and Morandi et al [7], under normal conditions there is no evidence of a naturally occuring space being extant at the dura-arachnoid junction. A space may appear at this point subsequent to pathological/traumatic processes that result in tissue damage with a cleaving along the structurally weakest plane in the meninges through the dural border cell layer. Furthermore, when a space does appear, it is not "subdural" in location but rather within a morphologically distinct cell layer. So according to works of these authors, the so-called spinal subdural hematoma could be viewed as a spinal dural border hematoma.

Spinal subdural hematoma usually presents with sudden local neck or back pain followed by neurological deficit. Sometimes the onset is chronic with no pain [4].

In our presented case, the clinical course of the disease could not be reliably ascertained. The patient has not reported acute pain. The child's deficit gradually worsened. This gradual delayed neurological deterioration, apparently occurs because the processes of primary hemostasis are only temporarily effective. Delayed bleeding has been reported to occur several hours or even days later [8]. Since the lumbar puncture level had been considerably below the lowest limit of the hematoma, the hematoma itself cannot be attributed to this procedure, but lumbar puncture may be one of the reasons for further neurological deterioration of the patient.

We obtained spinal MRI and CT scan on the operation day, which was six days post trauma. In his thoracic axial CT, the hematoma was hyperdense, circumferential and mainly anteriorly located. In the sagittal T2-weighted cervical MRI, the hematoma from C5 to midthoracic level, was hyperintense and mainly posteriorly located in the thoracic and thoracolumbar level. This is compatible with the timing of the imaging (6^{th} day after trauma and hemorrhage)[9,10] and is postulated to occur because of methemoglobin formation [11]. In the T1-weighted axial MRI scan of the lower thoracic region, areas of hyperintensity in the periphery of the hematoma were seen, suggesting methemoglobin formation. It has been said that visualization of the dura mater and preservation of the fat pad may favor of the intradural location of the hematoma [12]. Sagittal T2-weighted image of the lumbar region, showed mixed signals posteriorly.

Since the trauma to our patient was not of a degree to generally cause neural injuries, we can assume that the compressive effect of the hematoma was the main cause of neurological deterioration of the patient. In our case, a clear preoperative CSF study, the tension of the dura during operation and normal looking arachnoid and cord underlines the significance of compression as the pathophysiologic mechanism. We postulate that after initial control of the hemorrhage, minor rebleeding or expansion of the hematoma resulted in the deteriorating neurological status of the patient. Recent rebleeding as the main reason of neurological deterioration of our patient cannot be proved through our images.

The limited available literature mainly addresses the management of spinal epidural hematoma in the hemophiliacs [13–15,9]. Schmitz et al [16] has reported his survey of treatment and outcome in hemophilic patients with spinal epidural hematoma from 1977 to 1998. In his survey of eleven cases of such patients (4 hemophilia B and 7 hemophilia A), 3 were operated on and one of the three operated cases had complete recovery. The rest (8 patients), were conservatively managed with (75%) recovery. The cases are not completely comparable, but the author concluded that considering the fact that a significant number of the hemophilic patients suffering from intraspinal bleeding are children, and that there is a high risk of spinal deformity after decompressive laminectomy, conservative management should be preferred as much as possible. Although surgery has been associated with high morbidity and mortality, early surgical intervention is always indicated when the patient's neurological status progressively deteriorates. We operated on our patient after his neurological deterioration. Although our patient had been operated on late after the appearance of first neurological deficits, he recovered completely. This good functional outcome is not the rule. Review of the literature on this pathology indicates clearly that delay of surgical decompression for several hours after the appearance of an important sensorimotor deficit increases the risk of a poor functional outcome [17,18,7]. Considering his spinal level of neurological deterioration, we only approached the thoracolumbar hematoma. But the patient improved postoperatively and in his MRI taken 6 months later, the resorption of the hematoma in the cervicothoracic region is clearly seen. In fact the thoracolumbar subdural hematoma of this patient has been managed operatively and its cervicothoracic portion, conservatively.

Conclusion

This case calls attention to the clinical manifestation, radiological features and management options of the rarely reported intraspinal hematoma in the hemophilic children. Although this case has been operated upon for the hematoma in the thoracolumbar region, at the same time it can be considered a successful case of conservative management of intraspinal hematoma in the cervicothoracic region. Both conservative and surgical management could be an option in managing these patients considering their neurological course.

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