case report

Chronic spontaneous cervicothoracic epidural hematoma in an 8-month-old infant

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Spontaneous spinal epidural hematoma is an uncommon cause of cord compression in children, especially in infants. An 8-month-old infant was admitted to our hospital for a 40-day history of paraparesis in the lower extremities. This rapidly progressed to paraparesis with an inability to move the lower extremities. MRI of the cervicothoracic spine revealed an epidural mass with compression of the spinal cord. The infant underwent C7-T3 total laminectomies. The pathology and postoperative MRI confirmed spinal epidural hematoma from a vascular malformation. We present the case to highlight the significance of recognizing this chronic spontaneous spinal epidural hematoma and discuss the diagnosis, treatment options and prognosis.

Spontaneous spinal epidural hematoma is a relatively rare but disabling disease. Its incidence was found to account for 0.3% to 0.9% of all spinal epidural space-occupying lesions.[1] Spontaneous cervicothoracic epidural hematomas of the spinal canal during childhood are less rare. We report the rare case of spontaneous chronic cervicothoracic epidural hematoma in an infant. Our literature review revealed that this case is the youngest patient reported to have spinal cord compression secondary to spontaneous epidural hematoma. The hematoma and spinal cord compression were diagnosed by MRI, and the pathology was confirmed by postoperative histopathological analysis.

CASE

An 8-month-old infant was admitted to our hospital for a 40-day history of paraparesis in the lower extremities. This rapidly progressed to paraparesis with an inability to move the lower extremities. The initial symptoms were manifested as irritability and crying. His parents found that the infant's lower extremities were very stiff and hence sent him to the local hospital for treatment. During the treatment in the hospital, symptoms resolved spontaneously over a period of hours. Twenty days later, the patient rapidly progressed to paraparesis with an inability to move the lower extremities. The clinic presentations led the doctors to make an incorrect diagnosis of cerebral palsy.

Familial and past medical history was not significant for any major illnesses, congenital abnormalities or genetic syndromes. The infant had never undergone any invasive spinal procedures. Physical examination revealed normal upper extremity motion. There was tone in the left leg but no antigravity movement, and the right leg had only flicker movements. Muscular tone of the lower extremities was high. The sensory functions were impaired in all modalities evaluated. Because the patient was too young to act in concert with our doctors, we could not assess the sensory level accurately. Normal upper extremity reflexes were present. Patellar reflexes and deep tendon reflexes were active. Bilateral positive Babinski signs were present. All blood and coagulation tests (platelet counts, prothrombin time and partial thromboplastin time) showed normal values at that time.

Plain radiographs of the spine and chest were normal. MRI of the cervicothoracic spine revealed an epidural mass with compression of the spinal cord. On sagittal T1-weighted images and T2-weighted images (**Figures 1a, 1b**), the mass was epidural and within the spinal canal and spanned four vertebral segments (C7-T3). On axial images (**Figure 2**), the mass caused marked compression of the spinal cord, displacing it

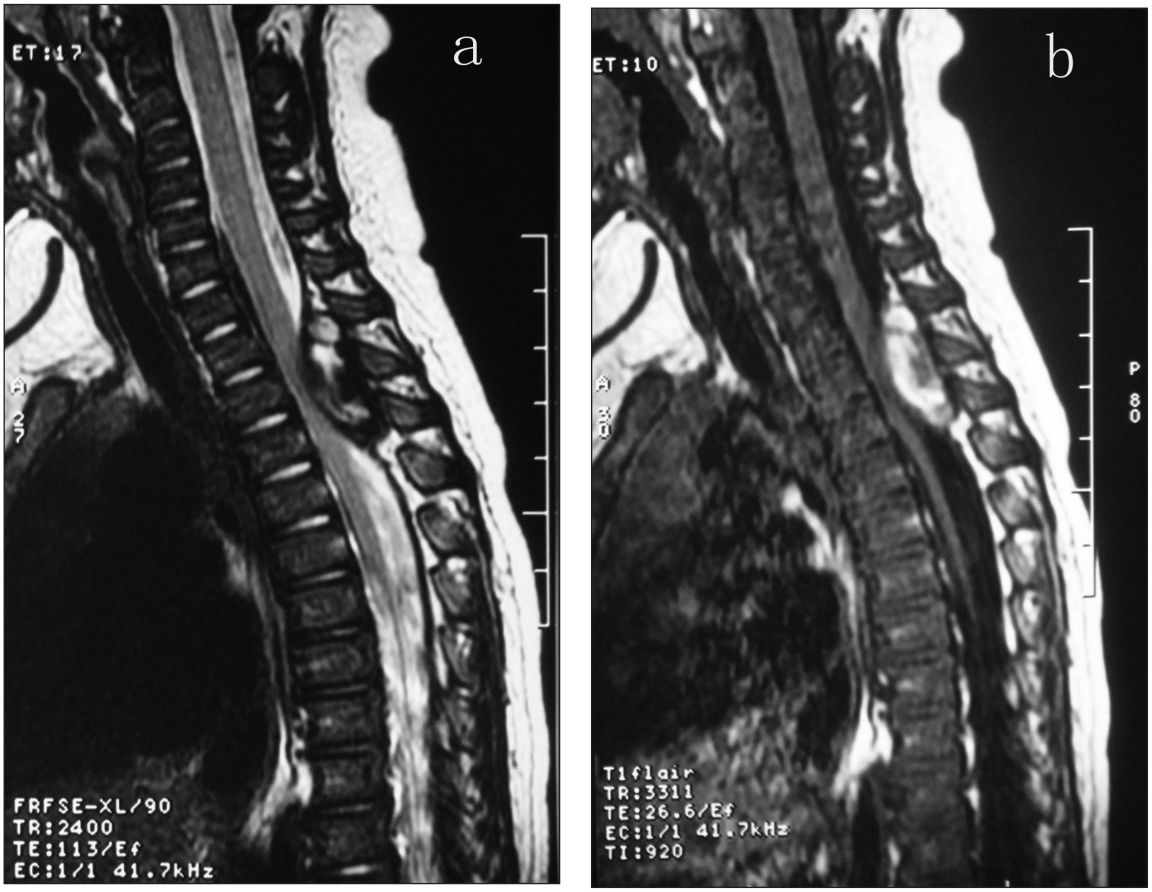
anteriorly and to the right. The mass appeared to be inhomogeneous. The presence of low-signal hemosiderin in the inhomogeneous mass revealed the epidural hematoma had formed some days previously. These findings were consistent with an epidural hematoma.

The infant underwent C7-T3 total laminectomies. Upon removal of the laminae, there was an angioma-like vascular mass on the dorsal surface of the spinal dura mater. After incision of the angioma-like vascular mass, there was an egress of liquefied and solid blood (**Figure 3a**). The mass had no vascular connection to the spinal cord and did not penetrate the dura mater. The lesion was totally resected from the dural surface. The epidural space was irrigated free of blood, and a small rubber catheter was passed cephalad and caudal to assure complete spinal cord decompression. The mass material was coated with blood, and no vessels could be grossly seen. Histopathological analysis of the mass revealed a vascular malformation (**Figure 3b**). Postoperative MRI of the cervicothoracic spine revealed extradural vascular malformation.

The patient had an uneventful postoperative course. The pain abated within the first 72 hours. Neurological examination revealed resolution of hyperreflexia, and he gradually regained strength, making a satisfactory recovery during the 20 days of follow-up. Four months after the C7-T3 total laminectomies, an MRI revealed no evidence of the compression of the spinal cord on serial imaging. The patient returned to live as a normal child with no significant symptoms and remains neurologically intact.

DISCUSSION

Spontaneous spinal epidural hematoma (SSEH) is an idiopathic accumulation of blood in the vertebral epidural space. It has been reported in all age groups, but it is most frequent after the fourth or fifth decade.[2] It is much less common in children, especially in infants. The most common site of SSEH in children, as recently documented in 27 cases, is C5-T1.[3] This was also the



Figures 1a,b. On sagittal T1-weighted images and T2-weighted images: the hyperintense mass spanned four vertebral segments (C7-T3), and appeared to be inhomogeneous. The presence of low signal hemosiderin revealed the chronic SEH.

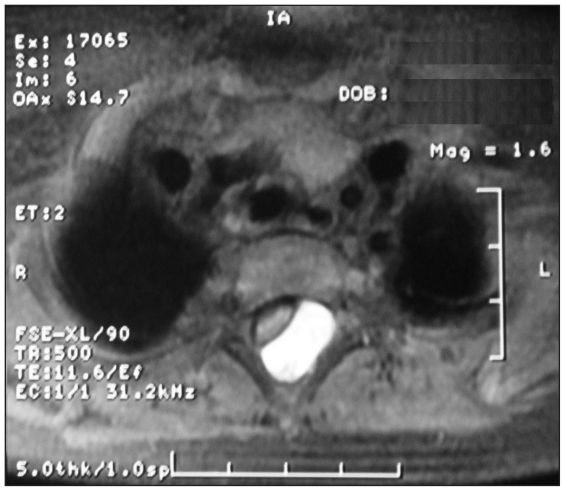


Figure 2. On axial images: The mass caused marked compression of the spinal cord, displacing it anteriorly and to the right.

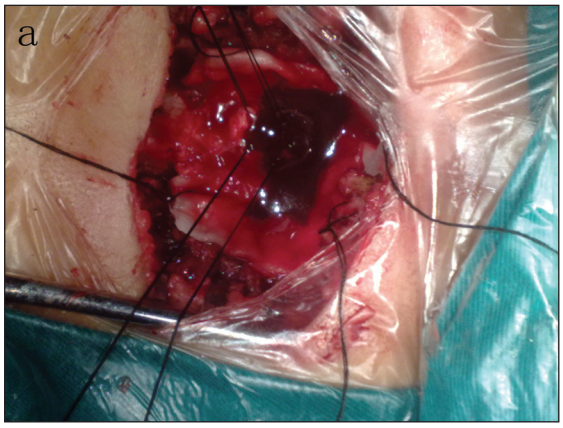


Figure 3a. After incision of the angioma-like vascular mass, there was an egress of liquefied and solid blood.

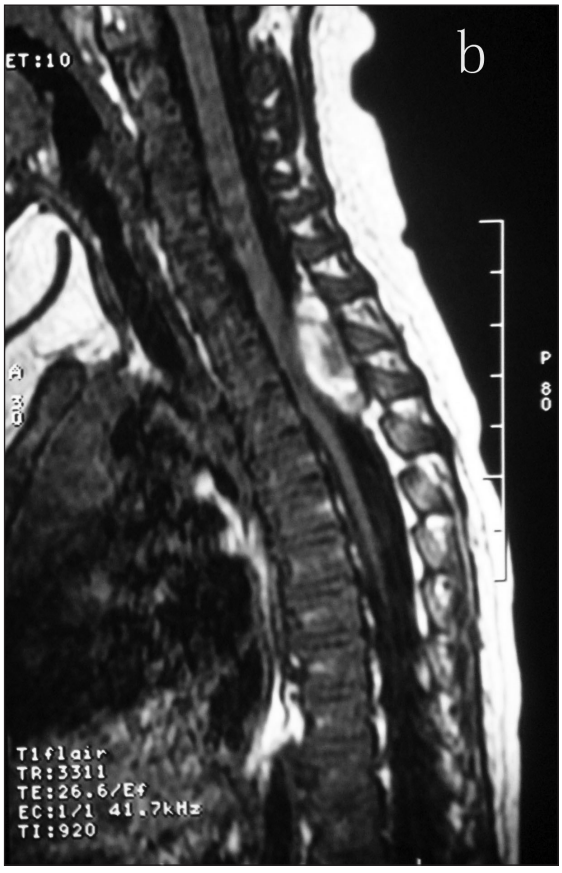


Figure 3b. Histopathological analysis of the mass revealed a vascular malformation.

level involved in our patient.

SSEHs can be acute or chronic. Chronic SSEH is defined as spinal compression for months or years. Compared to acute SSEHs, chronic forms are more rarely described and are generally located in the lumbar spine.[1,4] The rare chronic SSEHs of the cervical and thoracic spine described in the literature have a shorter progression of neurological symptoms,[5] probably due to the narrower spinal canal and the lower tolerance of the cervical and thoracic neurological structures to the space-occupying effect of an organizing hematoma.[6,7] Our patient had chronic spontaneous cervicothoracic epidural hematoma. The rare case was probably because our patient was unusual in that he was too young to verbalize complaints of the pain and the neurological deficits. The symptoms resolved spontaneously during the first treatment, and a suspicion of cerebral palsy delayed the correct diagnosis.

Predisposing factors for spinal epidural hematoma are found only in 50% to 60% of cases. Although often being suspected as a cause of spontaneous epidural hemorrhage, vascular malformations have rarely been demonstrated. In our patient, histopathological analysis of the mass and postoperative MRI of the cervicothoracic spine revealed a vascular malformation. The case may be an important piece of evidence that confirms vascular malformation as a cause of spontaneous epidural hemorrhage.

The clinical symptoms depend on the level of the hematoma.[2] The first symptom is usually pain, and it can either be localized to the level of the hematoma or be radicular in nature. In younger children, the initial symptoms can be nonspecific and can manifest as irritability and crying, especially in an infant.[3] Neurological deficits such as severe weakness and a change in sensory level may develop some hours or even days after the initial pain. Although our patient had similar initial symptoms and progression of neurological symptoms, he was

too young to verbalize complaints of the pain and the neurological deficits. Similar clinical presentations lead some doctors to suspect cerebral palsy. Without a correct diagnosis and early surgical intervention, our patient rapidly progressed to paraparesis with an inability to move the lower extremities.

MRI represents the diagnostic gold standard for SSEH.[7-9] The T1 and T2 intensity patterns are both equally helpful in discriminating against almost all other vertebral space-occupying lesions. Chronic epidural hematomas typically appear hyperintense in both T1- and T2-weighted images.[10] Due to the doctor suspecting cerebral palsy, our patient only had MRI of the skull before he was admitted to our hospital.

In general, the bleeding episode should be self-limited; and in most cases of acute spinal epidural hematomas, symptoms arise immediately in proportion to the size of the lesion and according to its location along the spinal cord. However, chronic spinal epidural hematomas could occur such that they produce few or no immediate symptoms or signs and even then go on to very slowly evolve over months.

Compared to acute SSEHs, in which surgical decompression must be carried out rapidly,[4,11,12] in the rarer chronic SSEHs, the time before decompression procedures are undertaken does not seem to be critical for the recovery of neurological function and, finally, a good clinical outcome. Coz et al[13] suggested that spreading of a hematoma remains possible until blood clotting has been completed. In most cases of chronic SSEHs, epidural hematoma frequently was small or moderate. Immediate conservative therapy can prevent progression of the hematoma, allowing for the regression of neurological signs and symptoms without operation.[14] However, in cases with repeated bleeding, one might expect a larger hematoma to occur, with serious and progressive neurological deterioration. The time that is available for reversal of the neurological deficit is limited, and surgical intervention is necessary before it is too late.[15] In our patient, satisfactory recovery was possible as children may have a better potential than adults for neurological recovery, and the patient received surgical intervention in time. A high index of suspicion can lead to an early diagnosis and intervention and hence to improvement of the overall prognosis.

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