Emerg Radiol (2008) 15:281–284
DOI 10.1007/s10140-007-0667-0

CASE REPORT

Spinal epidural hematoma in an infant as the initial presentation of severe hemophilia

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Received: 5 June 2007 / Accepted: 15 August 2007 / Published online: 18 September 2007

Abstract We describe the clinical manifestations, radiographic features, and management options of an extensive spontaneous spinal epidural hematoma in a 7-month boy who had severe hemophilia that had not been previously diagnosed, despite a baseline factor VIII level less than 1% of normal. We believe this to be the youngest reported case of a symptomatic spontaneous spinal epidural hemorrhage in an infant subsequently initially establishing a diagnosis of hemophilia.

Keywords Epidural hematoma · Hemophilia · Infant · MRI

Case report

A previously healthy afebrile 7-month-old boy was reported by a daycare provider to be fussy, irritable, and acting uncomfortable. He would not crawl and cried out with movement or change in position. On exam, he was uncomfortable with movement, particularly demonstrating significant pain with head movement. He was no longer able to sit up by himself, hold his balance when seated, pull himself up, or lift up his head. A local physician diagnosed a viral illness. Because he continued to be irritable, second and third opinions were sought.

Cervical spine radiographs suggested possible malalignment. Because of this and a family history of spine tumor, magnetic resonance imaging (MRI) was performed of the entire spine. This demonstrated an extensive, C2 through L4, partially enhancing epidural process (Figs. 1, 2 and 3) compressing the cord. The finding was confirmed on computed tomography (Fig. 4). This was reported as consistent with epidural hematoma. Abscess or tumors were considered to be less likely etiologies although not excluded. Surgical decompression was initially deferred. Nonaccidental trauma was briefly considered. The discrepancy between the extensive spinal epidural hematoma and the lack of trauma led to a hemostasis workup. This revealed a partial thromboplastin time elevated to 79 s. The international normalized ratio was normal. Further detailed family history eventually revealed that his maternal grandfather had hemophilia (and had died of human immunodeficiency virus infection acquired from a blood transfusion). However, this child, for reasons unknown, had never been worked up for hemophilia. Laboratory studies confirmed a severe factor VIII deficiency with factor VIII less than 1% of normal. This had never been previously tested. A new diagnosis of severe hemophilia A, factor VIII inhibitor was made in this otherwise healthy 7-month-old boy. The previously arranged computed tomographic-guided needle aspiration of the intraspinal "mass" was canceled. Factor VIII bolus followed by aggressive continuous infusion replacement therapy (547 U every 8 h) resulted in complete neurologic recovery. At 2 month follow-up, he was active, crawling, and moving well without irritability or pain.

Discussion

Nontraumatic spontaneous spinal epidural hematoma is uncommon in young children [1]. It is an extremely rare complication of hemophilia in infants and young children [2]. Infants with hemophilia are at a higher risk for this rare

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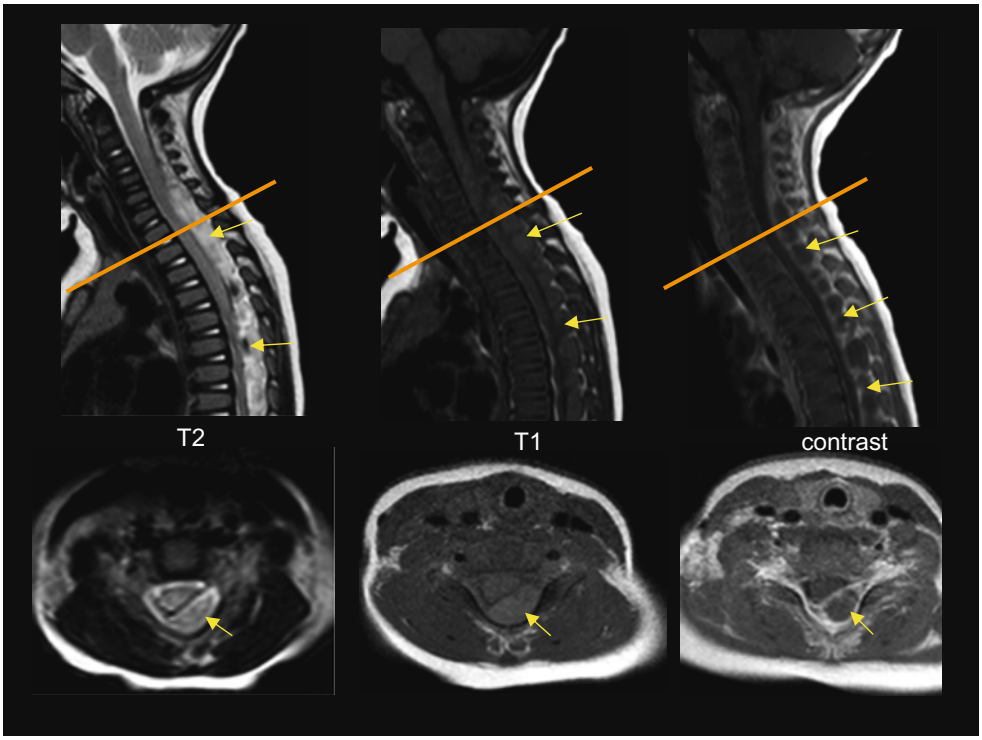

Fig. 1 Sagittal and axial (at cervicothoracic junction) MRI demonstrates an extensive T1 isointense, T2 hyperintense, peripherally enhancing lesion compressing the spinal cord

complication because of their mobility associated with frequent falls [3].

The imaging findings may mimic lymphoma, metastases, schwannoma, or abscess. The best distinguishing feature may be enhancement, which is very rare in spinal epidural hematoma [4]. When it does occur, such enhancement has been suggested to represent active bleeding (presumably of venous origin), contrast extravasation seen as central enhancement, or septa/vessels in extensions of the epidural fat seen as linear/peripheral enhancement [1].

Because spinal epidural hematoma can result in rapidly progressive and severe neurological deficits, it is a neurologic and potentially neurosurgical emergency. Prompt diagnosis is imperative. In adults, the classic triad of back

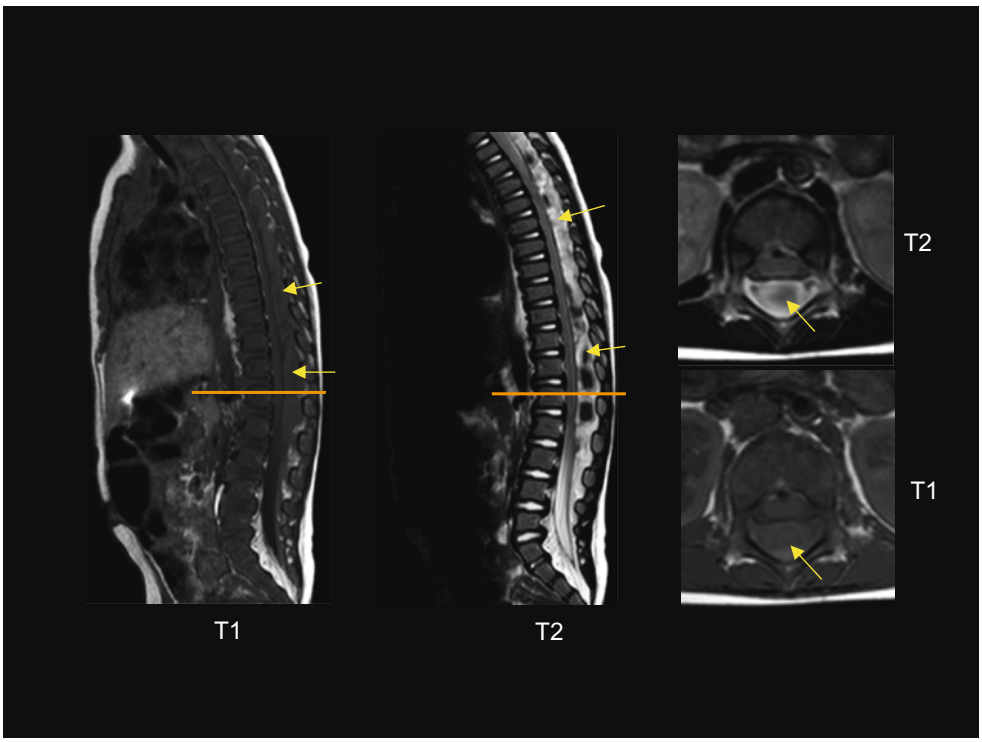

Fig. 2 Sagittal and axial (at conus level) MRI demonstrates T2 heterogeneity of the lesion

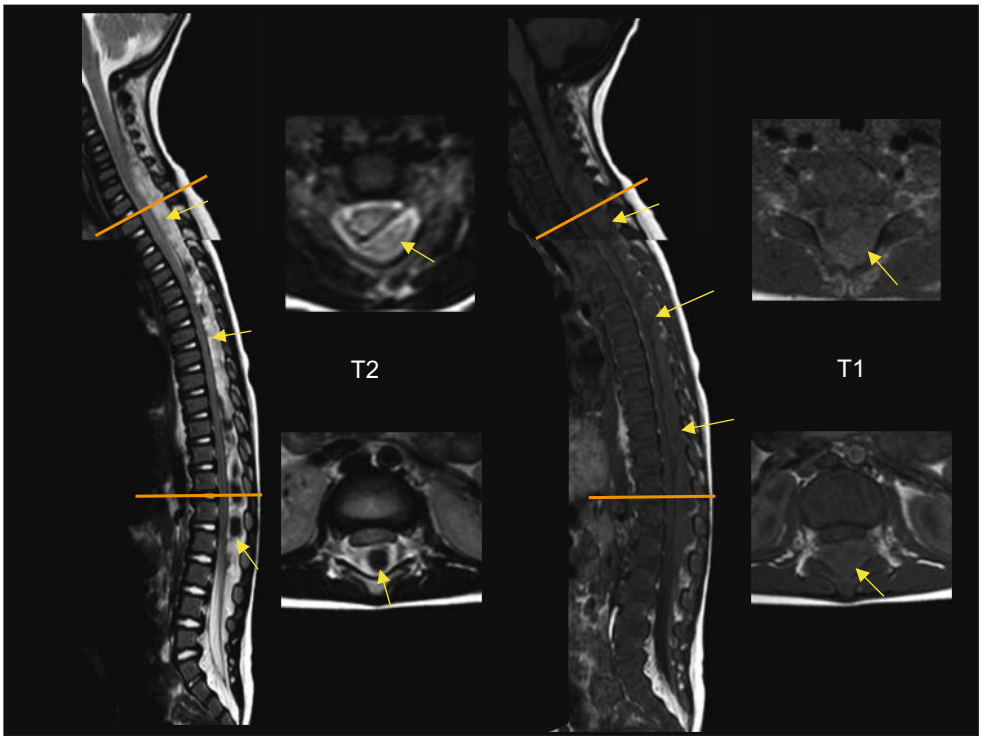

Fig. 3 Sagittal (entire spine) as well as cervicothoracic junction and conus axials reveal the heterogeneous T2 appearance

pain, radicular pain, and sensorimotor deficits can be used to establish the diagnosis of epidural hematoma [2]. However, in infants, the initial neurological findings may be subtle and nonspecific. Neurological assessments are often extremely limited in infants because they are not able to specifically localize or even express pain. Especially in an infant without a known diagnosis of hemophilia, this may result in a delay in diagnosis and treatment with a subsequently poorer prognosis. A high index of suspicion is required to diagnose a spinal hematoma in an infant without a known diagnosis of hemophilia, especially when there is no history of trauma or very minimal trauma. There is a

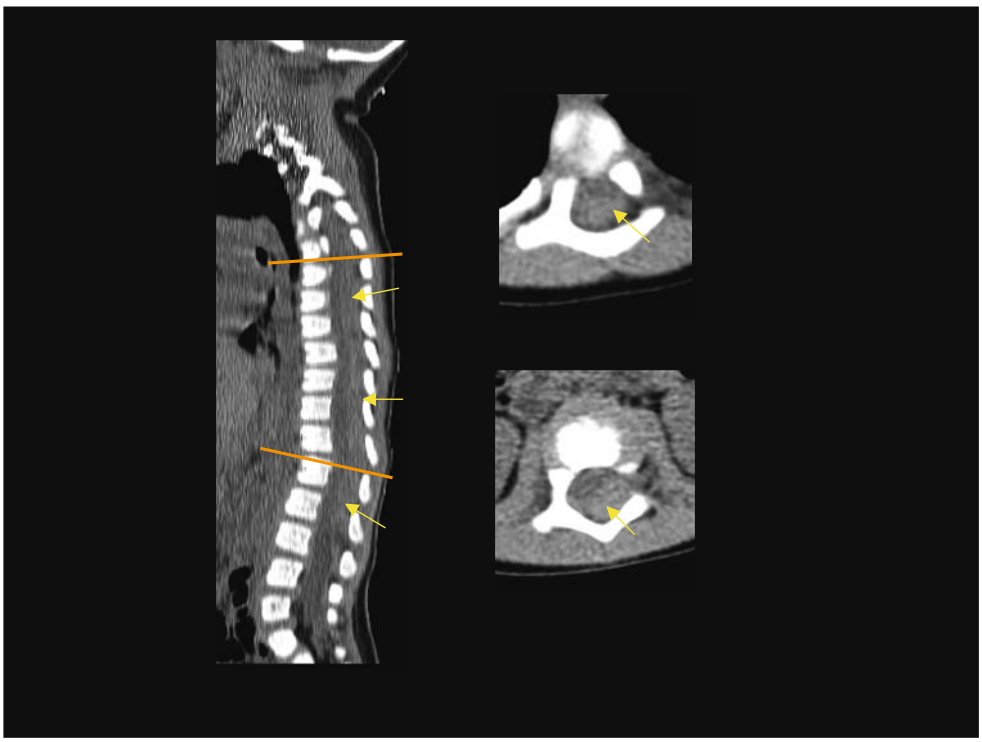

Fig. 4 Sagittal (entire spine) and axial (cervicothoracic junction and conus) CT demonstrates homogeneous hyperdensity compressing the spinal cord

propensity for spinal epidural hematoma in the cervical spine because of its greater mobility [5]. With cervical spine involvement, torticollis has been described as a sign that should prompt early MRI to avoid a delayed diagnosis [3]. Recovery depends on the duration and degree of cord compression, the severity of the deficit, and the rapidity of neurological deficit onset [4]. Despite an extensive spinal epidural hematoma, a complete neurological recovery is possible with conservative therapy if a prompt diagnosis is made, and there is aggressive immediate factor replacement and correction [5] This may avert the need for the potential high associated morbidity and spinal deformity of an extensive emergent surgical decompressive laminectomy.

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