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Case Report

Spinal epidural haematoma in a patient with haemophilia-B

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Spontaneous spinal epidural haematoma is rare. A case of haemophilia-B presenting with cervical spinal cord compression due to epidural haematoma, is reported. This is the second case in literature, to our knowledge, of intraspinal epidural haematoma in a patient with haemophilia-B. The importance of early diagnosis, especially with magnetic resonance imaging and surgical intervention, when deemed necessary, are emphasized.

Keywords: haemophilia; coagulopathy; spontaneous spinal epidural haematoma; magnetic resonance imaging

Introduction

Haemarthrosis and contractures are the most common musculoskeletal complications seen in haemophilia. Haemorrhagic complications of the nervous system are uncommon.[1,2] The most common neurological complication of haemophilia is bleeding into the peripheral nerve compartments and the incidence is between 1.3 and 20.4%.[3,4] Bleeding into the central nervous system is seen in about 2.2 to 7.8% of cases.[5,6] Intracranial haemorrhage remains as the leading cause of death, and carries a mortality of about 30%.[1,2,5,6] Spinal column and the spinal cord are rarely the sites of bleed.[7,8] The following case is reported because of the rarity of haemophilia B presenting with acute spinal cord compression and to show that major surgery can be undertaken in haemophiliacs without increased risk.

Case report

A twenty-six-year-old man presented with sudden onset of neck pain radiating to the right arm for 15 days. This was followed 3 days later by weakness of the right upper extremity and urinary hesitancy evolving over 48 h. There was no further progression. Two days prior to reaching our neurological services he developed progressive weakness of the other extremities; the lower followed by the left upper extremity, and urinary retention. Ten years earlier a bleeding diathesis after a bout of haematemesis had been diagnosed. Subsequently he had recurrent episodes of haematuria and haemoarthrosis of the left knee joint. General examination was unremarkable except for the left knee deformity. The vital signs were normal. There were no neurocutaneous markers. Neurological examination revealed normal higher mental functions, cranial nerves and ocular fundi. He had a grade (MRC) 2/5 spastic tetraparesis and impairment of all sensory modalities below C5 dermatome. Neck movements were restricted and tender. A clinical diagnosis of compressive cervical myelopathy was confirmed by MR imaging of the cervical spine that demonstrated an epidural lesion extending from C5 to T1 level. It was hyperintense on both T1 and T2 weighted images (Figure 1a and b). There was evidence of marked spinal cord compression from the posterior aspect.

The assays for clotting factors revealed a deficiency of factor XI and there was an impairment of coagulation functions. Hence, he received fresh frozen plasma and cryoprecipitates of factor XI preoperatively and an emergency decompressive laminectomy and evacuation of the haematoma was performed under coverage of coagulation factors' infusions. The epidural clot was semi-solid in consistency and there was no evdience of any vascular malformation. At the end of the procedure the dural sac regained normal pulsations. Postoperatively, the patient had shown good improvement in his neurological function and was ambulatory with support at the time of discharge. The replacement therapy for his coagulopathy was continued postoperatively.

Discussion

Spontaneous spinal epidural haematoma (SSEH) is a rare condition and was first described by Jackson[9] and Bain.[10] Patients with spinal epidural haematoma (SEDH) typically have an acute onset of severe

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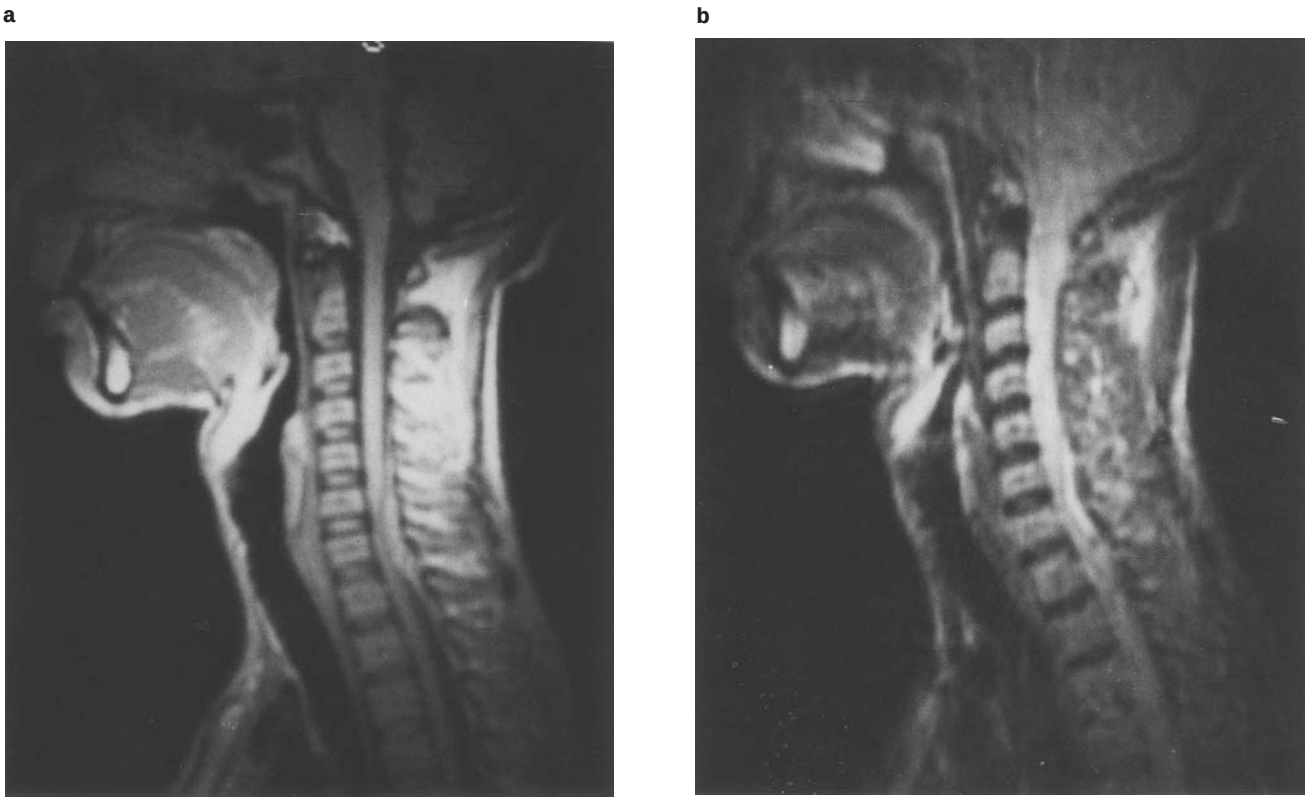

Figure 1 (a) Magnetic resonance imaging, cervical spine and upper thoracic spine, T-1 wt images showing the posteriorly located hyperintense mass with compression on thecal sac, maximum opposite to the C6 and C7 vertebral bodies. Note the straightening of the cervical spine secondary to the nuchalgia. (b) MRI (T-2 wt) The lesion remained hyperintense and the posterior epidural location well delineated by the thin hypointense interface between the dura and the mass (haematoma)

radiating neck pain or back ache followed by symptoms and signs of rapidly evolving root and/or spinal cord compression. Most common sites of bleeding are the lower cervical and thoraco-lumbar region in males and thoracic spine in females. MRI is the single most sensitive test in the diagnosis of spinal epidural haematoma. Serial MRI studies are useful in the progression of SSEH during conservative treatment.[11]

Trauma was the etiologic factor in about 10% of SSEH, in the report by Bruyn and Bosma.[12] In a review of 199 cases with SSEH by Rob *et al*,[13] no underlying cause could be identified in 52%; while bleeding diathesis accounted for 20.6% of cases. It was suggested that the posterior internal vertebral plexus plays an important role in these cases. SEDH is an uncommon complication of haemophilia. The exact incidence of SEDH in haemophilia is not known and review of the published literature revealed that the incidence is confined to occasional case reports. The first case of SEDH in a haemophiliac was reported by Tellagan and Ledaux as cited by Schenck.[14] Haemophilia-A accounts for a majority of these cases, probably because it constitutes 80–85% of haemophilia.[15] Only one case of SEDH in Haemophilia-B, has been reported so far in the literature.[11]

A good functional outcome with both operative and non-operative treatment modalities has been reported. Early decompressive laminectomy is advocated in patients with significant and rapidly progressive neurological deficit, to avoid the centromedullary myelomalacia that develops with sustained cord compression for more than 24 h.[12] In cases with complete neurological dysfunction, the results of surgical decompression may not be encouraging. The factors that determine a good functional recovery following surgical treatment include: an incomplete neurological deficit, a slow clinical progression especially with a long history of pain preceding the onset of myelopathy, involvement of short spinal segments, lumbar spinal location and early surgical intervention when deemed necessary.[16] The present case had a partial neurological dysfunction, a relatively slow progression, neck pain heralding the compressive myelopathy and a short segment of spinal cord compression. All these parameters serve as good prognostic indicators, and warrant an early operative intervention. However, in suitable patients an aggressive replacement therapy and expectant treatment even with long segment lesions are reported to yield good clinical results.[11,17,18]

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