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Case Report

Torticollis and right arm weakness due to atraumatic spinal epidural hematoma in a 5-year-old girl child

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ABSTRACT

Spontaneous spinal epidural hematoma (SEH) is a rare presentation as cause of neurological weakness in children and is neurological emergency. The most commonly affected site in children is cardiothoracic region. Early management is associated with better functional outcomes. We report a 5-year-old girl who presented with long standing neck pain now presenting to emergency with torticollis and right arm weakness around shoulder joint. Magnetic resonance imaging of C-spine revealed posterior epidural hematoma that extend from C3/C4 level down to C6/C7 level, it is causing moderate spinal canal stenosis and mass-effect on the spinal cord, with mild intramedullary abnormal T2 signal intensity indicating myelomalacia. There were no predisposing factors in our patient. Epidural hematoma was drained surgically and post-surgery patient started showing neurological recovery on follow. Cases of spontaneous SEH have been reported in infants as well and the clinical features in children are often nonspecific leading to a delay in diagnosis. This case report aims to raise awareness about this neurological emergency, in which early intervention is crucial.

Keywords: Torticollis, Right arm weakness, Spontaneous spinal epidural hematoma pediatric, Neurological emergency

INTRODUCTION

Spinal epidural hematoma (SEH) without history of trauma is rare in children. The term Spontaneous spinal epidural hematoma Pediatric (SSEP) was used to describe SEH without clear traumatic etiology in pediatric population. The condition is mostly observed in adults with a bimodal distribution with peak prevalence in the 2nd and 6th decades of life with an estimated incidence of 0.1 per 100,000 patients per year.[1-3] The clinical presentation of SEH without significant trauma in children is often nonspecific, including irritability (especially in reported cases of infant), neck pain, torticollis, and neurological deficit.[4-7] While in some cases, some form of cervical trauma precedes SEH.[8] Most cases manifest with acute onset pain at the level of the hematoma and sensorimotor deficit with or without bladder and/or intestinal disturbances.[2] Various etiologies, such as coagulation disorders and vascular malformations have been described.[9] The nonspecific presentation in children may lead to delayed diagnosis and treatment. Early surgical decompression likely leads to good outcomes.

CASE REPORT

A 5-year-old girl child with no previous history of health problems, presented to the pediatric emergency

department with sudden onset neck pain and torticollis 3 weeks back and progressed gradual, now child is having torticollis and right arm weakness and persistent pain in neck.

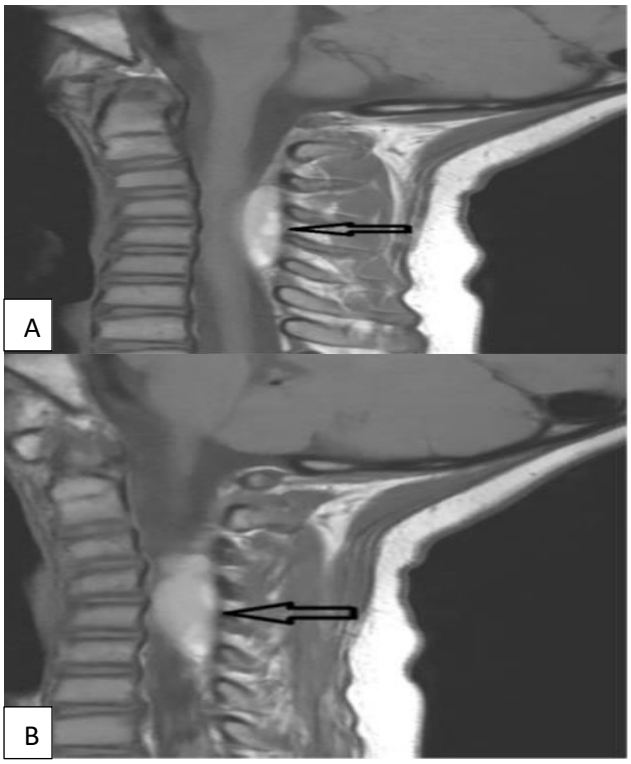

Figure 1 (A and B): MRI T1 images sagittal view C-spine subacute epidural hemorrhage.

No history of prior trauma was reported, no suspicion of non-accidental trauma. After 4 days the pain started to radiate to right shoulder, a muscular sprain was suspected in other hospital and discharged child on oral ibuprofen and topical diclofenac gel, which showed no improvement. No history of fever or abnormal body movements, or headache, the past medical history was unremarkable. Few days' later mother noticed right upper weakness, Persistent neck pain, neck stiffness, and torticollis leading to visit our emergency. Physical examination on admission showed an alert child with marked torticollis, pain-induced limitation of neck movements and tenderness on palpation of the neck muscles. Neurological examination was otherwise normal except right upper limb weakness around shoulder joint with power 3/5, elbow joint 1-2/5, Hand grip 4/5 sensation were intact and reflexes in right upper limb diminished. Emergency computed tomography (CT) done was normal and MRI of C-spine revealed posterior epidural hematoma that extend from C3/C4 level down to C6/C7 level, it is causing moderate spinal canal stenosis and mass-effect on the spinal cord, with mild intramedullary abnormal T2 signal intensity indicating myelomalacia (Figure 1 and 2, A and B). No vascular malformation was visible. MRI of the brain was normal. Conventional angiography was not performed; coagulation profile was normal. Due to the presence of mass effect on cord and clinical motor deficit in right arm, the patient was treated surgically with posterior cervical laminectomy and hematoma evacuation. Post-surgery after 3 weeks on follow in clinic her power in right upper limb had improved to grade 5/5.

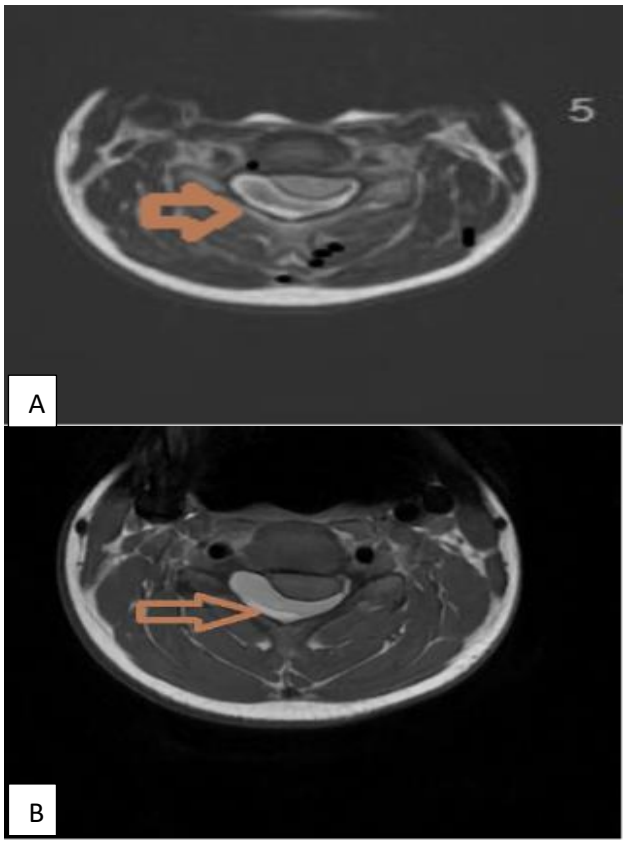

Figure 2 (A and B): MRI T2/ T1 images axial view C-spine subacute epidural hemorrhage.

DISCUSSION

Spontaneous epidural hematoma pediatric (SSEP) was first reported by Jackson in 1869 in a 14-year-old girl. A review revealed that SSEP s occur in all age categories and are by far the most common type of spinal hematoma.[2] Clinically spontaneous SSEP in children, may differ significantly from those in adults. In adults there is a clear male preponderance, whereas in children no such sex difference has been reported.[10] Most of the hematomas are located in the thoracolumbar region in adults, whereas involvement of the cardiothoracic region, usually C5-T1, is more common in children. The clinical presentation depends on the size of hematoma, location, and volume as estimated by the extent of spinal segments involved and duration since onset. In children, the initial clinical signs and symptoms are often nonspecific and include irritability in infants, neck pain, rigidity or abnormal posture, torticollis gait abnormality, limb weakness. Our patient had involvement of C3-C7, maximum curvature of the cervical spine, increased cervical spine mobility, probably explains the propensity of the cervical spine to be involved. Bleeding in

spontaneous SSEP is thought to result from rupture of the valve less epidural venous system. The internal vertebral venous plexus is divided into anterior and posterior parts.[11] The anterior part is closely attached to the posterior longitudinal ligament by firm connective tissue strands called Hoffman ligaments and is stable. The posterior part courses loosely through the epidural fat, which extends laterally to surround the nerve roots. The sudden elevation of intrathoracic or intra-abdominal pressure induced by activities like crying, coughing, straining or trauma can cause rapid increase in backflow into this valve less venous system, Hence, most spontaneous SEPs are located in the posterior aspect of the spinal canal and may extend into lateral gutters.[12] Hematoma in our patient was posterior aspect to the cervical cord.

Our patient had prominent torticollis and persistent neck pain and right upper limb motor weakness, probably because of mass effect, in children, early clinical features are nonspecific, often leading to a delay in diagnosis. Numerous different disorders present with acute-onset limb weakness, these include, acute disseminated encephalomyelitis, immune myelitis, Guillain-Barre syndrome. Non accidental trauma can have varied presentation and should always be excluded. Clinical assessment, CSF analysis, imaging of brain and spine, and nerve conduction studies and needle electromyography include the initial workup. Risk factors for spontaneous SEP include coagulation disorders, vascular malformations like venous angiomas, hemangiomas, or epidural varices and whooping cough Iatrogenic cases have been documented after lumbar puncture and epidural anesthesia. The hematoma causing mass effect needs surgical management and the determinants of neurological outcome depends on duration from onset of clinical features to surgery and the severity of neurological deficits at the time of presentation. Appropriate time for surgical decompression from symptom onset has been described as within 12 to within 48 hours in adults but most of the pediatric cases present later. The time from symptom onset to surgery has ranged from 2 to 14 days in children. In our case it was more delayed (more than four weeks). laminectomy of the involved segments is considered the surgery of choice. However, there is a concern that laminectomy might limit normal development of the spinal column in children and may result in kyphosis, scoliosis, or tethered cord. Therefore, some authors have advocated laminotomy for decompression in children.

CONCLUSION

This case report highlights spontaneous SEP is rare in pediatrics and to encounter within emergency, but high suspicion should be kept in child presenting with torticollis and neck pain with motor weakness. It is a neurosurgical emergency, and early intervention is necessary.

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