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ORIGINAL ARTICLE

Spontaneous epidural spinal haematoma in children caused by vascular malformations

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Abstract
Purpose The occurrence of spinal epidural haematoma of 'spontaneous' origin in adults is a well-documented entity, though it is rare in children. In the literature to date, there are few cases of this kind of spontaneous haematoma proven to be due to an underlying vascular abnormality.
Method Retrospective review of two cases of children under 15 years of age with spontaneous epidural spinal haematoma.
Results Underlying arteriovenous malformations were identified in both cases. Intra-operative photographs and histological sections of these anomalies are presented.
Conclusion These are the first two such cases described with clinico-pathological correlation.

Keywords Spinal · Epidural · Haematoma · Arteriovenous · Malformation · Children

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Introduction

Spontaneous epidural spinal haematoma (SESH) is a rare cause of spinal cord compression in children [1]. Although well recognised in adults, reports of this entity in the paediatric population are few and are ascribed to idiopathic causation. We present two children with their surgical management, outcomes and pathological features.

Case 1

A previously healthy 8-year-old girl developed severe neck pain while dancing. She also reported feeling a 'pop' as the pain developed. Over a 6-week period, she became increasingly lethargic with progressively worsening mobility. After a further week, she attended the casualty department of the local district general hospital unable to walk.

Clinical examination on admission revealed MRC grade 3/5 spastic tetraparesis and urinary retention. Spinal magnetic resonance imaging (MRI) revealed an epidural lesion compressing the theca on the right from C4/5 to C7/T1 levels (Fig. 1). The lesion was centrally bright on T1-weighted imaging with a peripheral lower signal intensity rim. The signal characteristics of this lesion were consistent with an acute or subacute epidural haematoma. No other abnormality was seen in the neuroaxis. All haematological investigations on admission were within normal limits.

The girl was taken for an urgent C5–C7 laminoplasty and evacuation of haematoma. Significant bleeding was encountered from abnormally dilated arterialised epidural veins, which was quickly controlled by diathermy. These veins extended caudal to the laminotomy (Fig. 2), but as no vascular imaging was available to provide architectural

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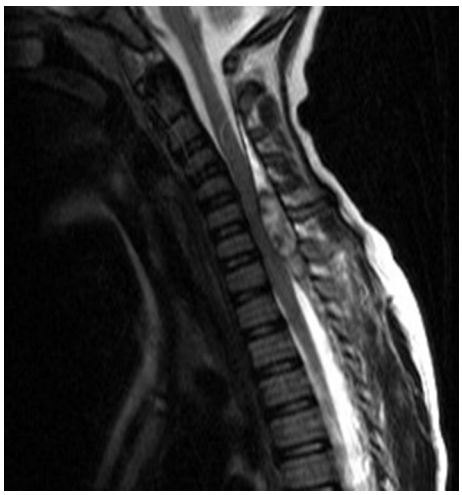

Fig. 1 T2-weighted image of cervico-thoracic spine illustrating dorsal epidural lesion

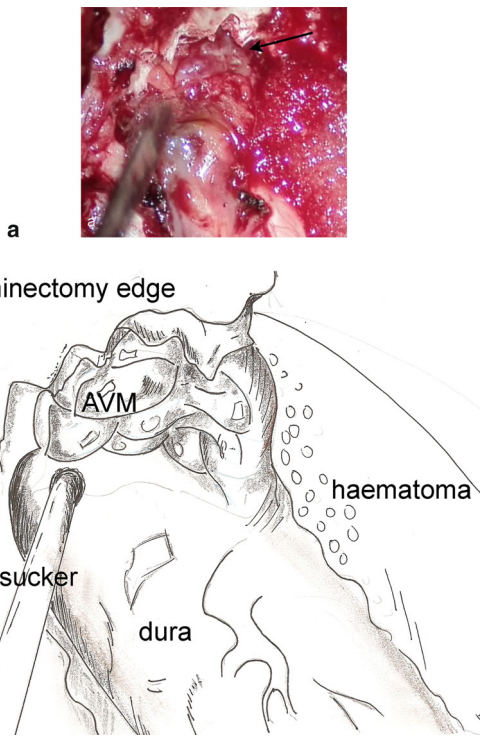

Fig. 2 Intra-operative photograph (**a**) demonstrating residual arteriovenous malformation underneath cervical lamina (*arrow*) with accompanying illustration (**b**) depicting arteriovenous malformation (*AVM*) under cut edge of lamina, with dura and overlying haematoma exposed

information (on what we believed to be an arteriovenous malformation), we elected not to extend the exposure within the immature spine. With the existing exposure, the abnormal vessels were removed from the dural sac and excellent haemostasis achieved. Once the veins at the level

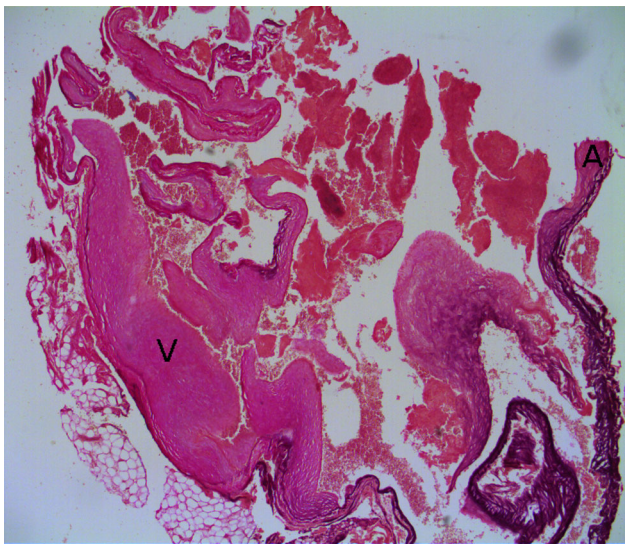

Fig. 3 Histology showing vessel walls with varying thickness including a thin arterialized vessel wall (*A*) and a thick fibrous walled vessel (*V*) (elastic Van Gieson stain, ×20)

of exposure had been excised, the bleeding from the remaining veins was evidently no longer arterial.

Post-operatively, the child awoke immediately to make a good neurological recovery over the subsequent 3 weeks. At discharge, she was ambulant independently with normal bladder function. Time-resolved imaging of contrast kinetics (TRICKS) sequence MRI scan performed post-operatively did not show any residual haematoma or abnormal vasculature.

Histology revealed blood clot and a complex arrangement of vessels with varying wall thickness, including venous like structures but with fibrous thickening and some arterialized vessels with thinner walls (Fig. 3). The features were those of a vascular malformation, consistent with an AVM.

Case 2

An otherwise healthy 13-year-old boy developed sudden-onset interscapular pain with no obvious precipitating cause. The following day, he developed progressively severe lower limb weakness and urinary retention. He was admitted to a local children's hospital where neurological examination revealed paraplegia (MRC grade 0/5) with areflexia, loss of anal tone and a T4 sensory level. MRI of the thoracic spine (Fig. 4) demonstrated a mixed-signal epidural lesion located posteriorly and to the left extending between C7/T1 and T4/5. The lesion displaced the spinal cord anteriorly and its signal characteristics were most in keeping with an acute or subacute epidural haematoma.

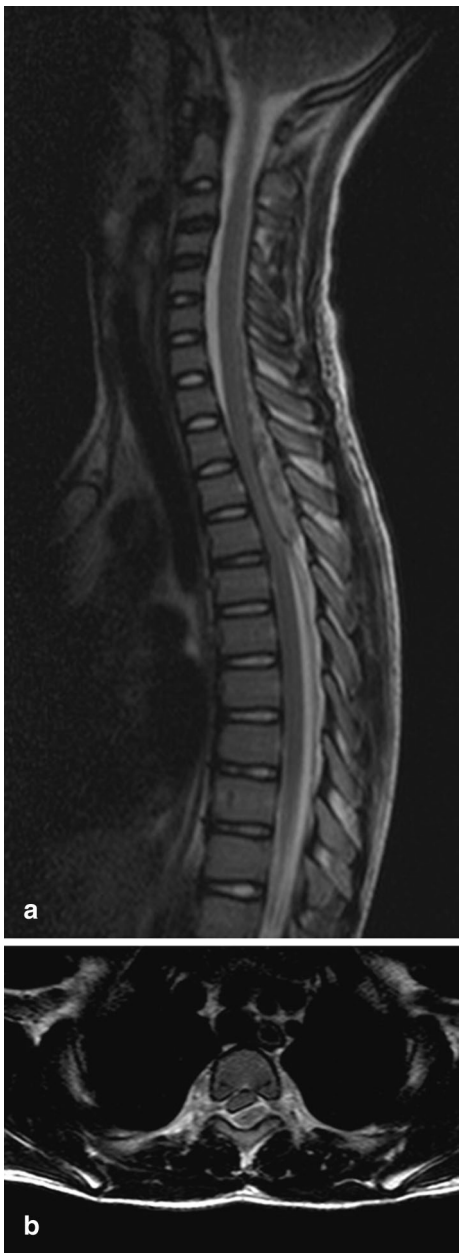

Fig. 4 Sagittal (**a**) and axial (**b**) T2-weighted images demonstrating epidural lesion displacing spinal cord anteriorly

He underwent emergency T1–T5 laminoplasty and evacuation of an epidural haematoma and resection of an AVM. Histological analysis revealed fragments of haemorrhagic tissue containing vascular channels with abnormally thick and irregular walls, which were also, in places, very thin (Fig. 5). These findings were consistent with the presence of an arteriovenous malformation.

Post-operatively, this boy's neurological deficit had improved to MRC grade 4/5. At the time of discharge from hospital, he still experienced intermittent urinary incontinence but was able to walk without assistance. By 3 months post-operatively, he was independently ambulant with very mild right lower extremity weakness on close scrutiny of his gait, though his intermittent urinary continence persisted. MRI of his thoracic spine performed at that time demonstrated no flow voids or abnormal contrast enhancement pattern to indicate residual vascular malformation.

Discussion

Spontaneous epidural spinal haematoma (SESH) is an uncommon cause of spinal cord compression in children, with 37 cases previously reported in English literature to date. The clinical presentation may be variable or even non-specific making it difficult to diagnose. The primary complaint is usually spinal pain at the level of haematoma, which may or may not be accompanied by radicular pain appropriate to that level [2, 3]. In some cases, irritability may also be the only early presenting feature of SESH. Cakir et al. [4] describe a case of a 9-year-old girl with a spontaneous thoracic epidural haematoma initially misdiagnosed as Guillain–Barré syndrome. Intravenous immunoglobulin therapy did not improve her symptoms, prompting MR imaging which revealed a thoracic epidural haematoma. This case illustrated the importance of adequate, timely imaging in supporting the diagnosis.

Magnetic resonance imaging is considered to be the gold standard in diagnosing spinal epidural haematomas [3, 5]. Not only does MRI help to identify haematoma, but it also helps to estimate its age and may help to identify the presence of an underlying vascular malformation. Furthermore, TRICKS MRI provides high-resolution vascular imaging of the spine and is particularly invaluable when studying arteriovenous malformations. TRICKS MRI thus obviates the need for digital subtraction angiography, which is considerably more invasive and technically demanding.

In our first case, the child had a 5-week interval between the onset of symptoms and the development of a neurological deficit. Patel et al. [2] describe patients primarily complaining of pain ranging from several hours to several days prior to the development of neurological signs. Interestingly, Matsumae et al. [6] describe a case of a 13-year-old boy who developed spastic paraparesis which was managed non-operatively as plain C-spine radiography and CSF analysis were normal. Symptoms completely resolved within a fortnight only to recur over 3 years later, when myelography was performed revealing an epidural mass lesion extending from C4 to C7 vertebral levels. Subsequently laminectomy was performed to remove the haematoma and intra-operative inspection did not reveal the presence of any abnormal vessels or neoplastic process.

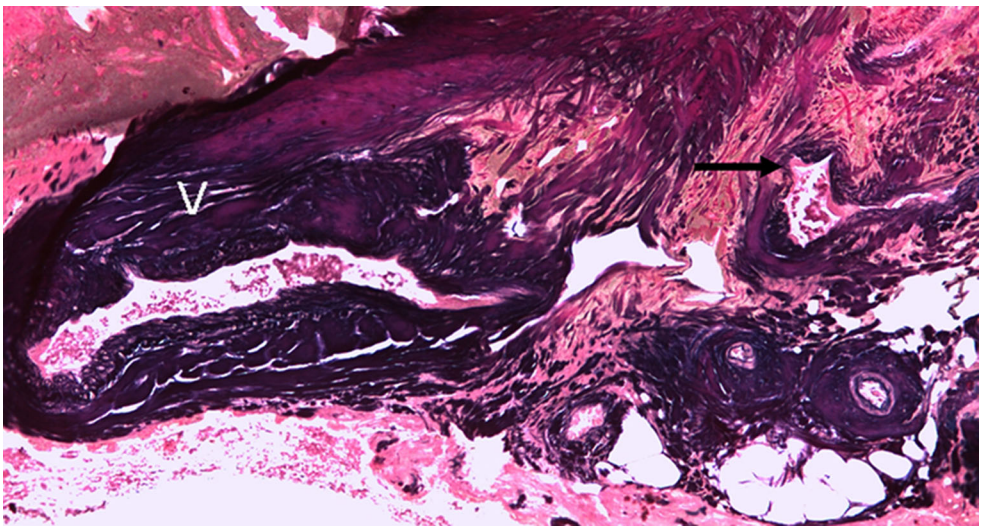

Fig. 5 Ruptured arteriovenous malformation surrounded by extensive haemorrhages. *Thick*, irregular blood vessel (*V*) and another vascular channel revealed segmentally thinned elastic laminae (*arrow*), showing transition between artery and vein (elastic Van Gieson stain, ×20)

Abram et al. [7] report a 10-year-old girl with three discrete separate episodes of paraparesis, with MR imaging interpreted as unremarkable after the first two occasions. After the third episode, MRI displayed a cervico-thoracic junction epidural haematoma. Emergency decompressive laminectomy and clot evacuation was performed with no AVM identified. Interestingly, spinal angiography on that admission was also interpreted as unremarkable and she was discharged. Follow-up spinal angiography at 2 years was suggestive of a vascular abnormality. However, exploratory laminectomy did not reveal evidence of an AVM.

The site of epidural haematoma in children is most commonly dorsal [8] and occurs largely between C5 and T1 vertebral levels. The underlying abnormality or precipitating cause of the haematoma is found in only 50–60 % of cases [9], though the bleeding is generally thought to be venous in origin. Normal activities of daily living may be responsible and cases of SESH have been reported after activities such as sneezing, coughing and bending over. These activities may cause changes in intra-abdominal or intra-thoracic pressure altering the dynamics of blood flow within the veins of the valveless epidural venous plexus [2, 8]. Other possible aetiologies include coagulopathies, AVMs or an underlying neoplasm. In many cases, an underlying abnormality, whether anatomical or physiological, may never be discovered [10].

The presence of abnormal epidural veins identified intra-operatively was a feature our first case. Akutsu et al. [11] describe a similar occurrence in a 27-year-old man with a SESH whereby, after evacuation of the haematoma, the presence of abnormally dilated epidural veins was discovered. These veins were found to form an epidural plexus and histological assessment showed varying calibre of the vessels with muscular hyperplasia and collagenous tissue contributing to their thickening. Some of the veins even had an internal elastic lamina like an artery, but no arterio-venous anastomosis was identified.

The presence of an AVM identified as the underlying cause of SESH is rarer still, comprising 2 % of all spinal lesions in children below 10 years of age [12]. Chen et al. describe a case of an 8-year-old boy who developed back pain radiating to the right anterior chest wall and became paraplegic. Investigations showed an epidural lesion and the operative finding was an AVM fed by a radicular artery from the right T1–T2 root entry zone. However, in most cases, an underlying vascular malformation as the cause for SESH may never be found. Tewari et al. [9] reports a case series of 3 children (aged 11, 8 and 5 years) each with spontaneous epidural haematomas where no underlying vascular malformation was identified.

The role of spinal digital subtraction angiography (DSA) in the investigation of spinal epidural haematomas is reserved for those cases where vascular malformations are suggested on MR imaging [1, 13]. Interestingly, Abram et al. [7] report a case in a 10-year-old girl where DSA was performed 8 days post-operatively and revealed no spinal vascular abnormality. However, a repeat angiogram was performed at 2 years was suggestive of 'an unusual vascular abnormality' in the right-sided extradural space at C7 level. The patient then underwent subsequent exploratory laminectomy, but operative findings revealed no vascular abnormality.

In the case of a 14-year-old girl described by Rosenberg [8], spinal DSA was performed at 2 weeks after the patient was discharged from hospital. No spinal vascular

abnormality was identified in this study. Other than the two children mentioned, no cases of spontaneous spinal epidural haematoma in the available literature underwent spinal angiography.

The presence of a vascular malformation might be anticipated in a child presenting with a spontaneous epidural haematoma even without overt evidence of flow voids on MRI scan. There is a potential for significant arterial blood loss and the evacuation of such a haematoma will be safest in the hands of a suitably experienced surgeon. Thirty-seven cases of SESH have been reported in the world literature. These two cases are the first to present with clinico-pathological correlation. The optimal clinical follow-up for such cases remains to be defined as does the natural history of asymptomatic lesions.

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Conflict of interest None.

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