CASE REPORT

Cervical spine arterio venous malformation

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SUMMARY

We present an unusual case of a young patient who presented to our emergency department with a history of sudden onset of pain on the left side of the neck and numbness to the left arm after lifting a small weight. Patient continued to work as normal for approximately 30 min after the event and then attended the emergency department as numbness in the left arm was not resolving. On examination there was no sensorymotor deficit in the lower limbs but neurological deficits were found in the upper limbs which made us suspect a diagnosis of cervical spine injury/brachial plexus. The patient then rapidly developed numbness in both upper and lower limbs and eventually became aphasic and developed a rapid, shallow respiration and was unable to maintain the airway. The patient was then intubated and ventilated. The patient was then transferred to a neurosurgical centre after the relevant investigations was found to have an arteriovenous malformation of spinal cord.

BACKGROUND

We draw the attention of emergency medicine physicians to those patients presenting to the department with a sudden onset of neurological deficits against the background of trivial neck trauma to bear in mind among other causes arteriovenous malformations of spinal cord and the rapid deterioration that these patients could potentially suffer if the relevant investigations and referrals are not undertaken.

CASE PRESENTATION

We present an unusual case of a young patient who presented to our emergency department with a history of pain in the left side of the neck and numbness to the left arm after lifting a small weight using both upper limbs.

The patient complained of having felt a crack in the neck when leaning forward after removing a 'tack' from a horse and then developed numbness of the left arm soon afterwards.

The patient continued to work as normal for approximately 30 min after the event and then attended the emergency department as numbness in the left arm was not resolving.

On arrival at the emergency department the patient was alert, Glasgow Coma Scale was 15. The pulse rate was 85/min, temperature 37.2, blood pressure was 130/70 mm Hg and respiratory rate and oxygen saturation were within normal limits. On examination, there was no sensory deficit in the lower limbs but left upper limb had paresthesia on C-5 to T-1 dermatomal distributions and motor functions were 4/5(MRC grading) in elbow/wrist/

hand muscles. Right upper limb had paresthesia on C-5 to T-1 dermatomal distributions with 5/5 motor functions. On history and physical examination we arrived at a tentative diagnosis of cervical spine injury/brachial plexus injury. All precautions of cervical spine injury were taken and performed the routine investigation for above diagnosis.

While patient was waiting for the investigations there was rapidly progressive weakness and numbness in both upper and lower limbs and eventually became aphasic and developed a rapid, shallow respiration and was unable to maintain the airway. A decision was then made to intubate and ventilate. Because of this rapid deterioration we thought of investigating the patient to exclude the possibilities of cerebrovascular incident or an acute intracranial haemorrhage.

INVESTIGATIONS

We drew blood for full blood count, urea and electrolytes, prothombin time and International Normalised Ration, random blood sugar which were all within normal limits.

Urgent CT scan of the brain and cervical spine which was reported as normal and excluded the possibilities of cervical spine injury and intracerebral haemorrhage. But still it could be a case of SCIWORA (spinal cord injury without a radiological abnormality). At the same time there was a suspicion of a vascular event involving the upper spinal cord and the brain stem. Because of the neurological deterioration we discussed with neurosurgical referral centre and patient was urgently transferred to the regional neurosurgical centre for further evaluation and management.

At the neurosurgical centre a CT head, CT neck, CT angiogram (from aortic arch to circle of Willis) were reported as normal. MRI of the brain and spine (figures 1 and 2) was suggestive of a dural venous fistula or cord arteriovenous malformation (AVM). Catheter angiogram (figure 3) was done and reported as

1. Spinal cord AVM at C-1/C-2
2. Arterial supply from the muscular branches of vertebral artery bilaterally, and anterior
3. Spinal artery(ASA)
4. ASA origin from distal right vertebral artery
5. Pseudoaneurysm

The patient was then transferred to the neurosurgical ICU and weaned off the sedation.

On further neurological evaluation, the patient remained quadriplegic but had no evidence of cognitive impairment but was dependent on the ventilator.

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Figure 1 C-2 level cord haemorrhage.

DIFFERENTIAL DIAGNOSIS

- ▶ Cervical spine injury
- ▶ Brachial plexus injury
- ▶ Intracerebral haemorrhage

TREATMENT

On arrival to the emergency department adequate analgesics were given. When the patient deteriorated, the patient was intubated and ventilated with peripheral intravenous access. Patient resuscitation was done and necessary supportive care was given as for a ventilating patient.

At the neurosurgical centre decision was made as for conservative management because surgical treatment was not indicated at this stage. The plan was to re-scan in 6 weeks' time.

Antinuclear antibody, cryoglobulin studies were performed to exclude other pathologies like systemic lupus erythematosus and Wegener's granulomatosis.

Figure 2 C-1/C-2 spinal cord haemorrhage.

Figure 3 Catheter angiogram—AVM at C-1/ C-2 level.

Due supportive care was given to the patient and tracheostomy was eventually performed to support the airway and breathing. Later patient was eventually weaned off the ventilator and a speaking valve was inserted to facilitate the communication. There was recovery of some sensory and motor function with increasing tactile sensation in the left upper and lower limbs and was able to move the right index finger voluntarily in about 2 weeks postinjury.

OUTCOME AND FOLLOW-UP

The patient was discharged from the neurosurgical unit and transferred to the spinal rehabilitation centre after 4 weeks of injury and is being followed up on a monthly basis at the neurosurgical hospital.

DISCUSSION

Vascular malformations of the spine and spinal cord are considered uncommon lesions, representing 3–16% of all spinal space-occupying lesions. Dural arterio venous fistula (AVF) is more common than spinal cord AVMs.¹ Spinal cord AVMs usually present in the third decade of life, but they can be diagnosed in children <16 years old in 20% of the cases.² In the paediatric population male predominance is shown, but no sex predominance is found in the adult population.^{3 4}

The first clinical description of a spinal vascular malformation was in 1890 by Berenbruch,⁴ who did not recognise the lesion as a vascular entity at surgery but only later at autopsy. Two decades later, Krause⁴ recognised a spinal vascular malformation at surgery.

The introduction of selective spinal angiography in the late 1960s and early 1970s further improved the anatomical and haemodynamic understanding of those rare lesions. Multiple eponyms and classifications have been used for the various types of vascular malformations of the spine and spinal cord and several classifications have been proposed.^{5–7} Spinal AVMs are divided into subgroups based on morphological data, angiographic interpretations and the relationship of the shunt with anatomical compartments of the spinal cord. The classifications assume that the lesions are static in time and are not dynamic.⁸

Spinal arteriovenous malformations have been divided into dural (type 1), intramedullary glomus (type 2), juvenile (type 3) and perimedullary direct arteriovenous fistulae (type 4), type 4 has been further subdivided into three subtypes simple fistulae fed by a single arterial branch (type 4a), intermediate sized fistulae with multiarterial feeders (type 4b) and giant multipe-dunculated fistulae type 4c.⁸⁻⁹ Syndromes associated with spinal vascular malformations are also included in this group, such as Klippel-Trenaunay, Parkes-Weber syndromes and Foix-Alajouanine syndrome.⁷

Learning points

- ▶ Low threshold to think about arteriovenous malformation of spinal cord in patients who present with sudden onset of numbness with a trivial trauma.
- ▶ Monitor clinically the progress and prepare for emergent intubation and ventilation as such patients can have cord infarction and such could deteriorate rapidly.
- ▶ Exclude the possibility of intracranial haemorrhage even in such presentations as this has a bearing on the prognosis.
- ▶ Early referral to neurosurgery in such cases.

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Competing interests None.

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