CASE REPORT

A giant spinal arterial aneurysm in a child presenting as quadriparesis

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SUMMARY

A giant spinal aneurysm from anterior spinal artery not associated with arteriovenous (AV) malformations is unusual and no such cases have been reported in children. Few cases have been described as part of AV malformation complex and coarctation of the aorta. We report a case of anterior spinal aneurysm in a 1-year-old girl causing a subarachnoid haemorrhage and a cervical cord lesion. The diagnosis was confirmed with a multislice CT angiography. A microsurgical decompression was performed and excision of aneurysm was unsuccessful but neurological deficits were improved. No further approach was accepted by the parents. The mechanism for the development of spinal isolated aneurysms is not clear; it can be related to congenital vessel abnormalities and genetic origin. The multislice CT angiography is a very useful method to demonstrate the features of this entity. Previous reports of isolated spinal aneurysm are reviewed.

BACKGROUND

The purpose of this paper was to present the unusual case of this entity in a child and discover further considerations in this case.

CASE PRESENTATION

The present case was a previously healthy 1-year-old girl who presented to the emergency room of the local hospital because she developed a sudden onset of quadriparesis state and neck pain over an 8 h period. Physical examination disclosed superficial breathing with marked use of the auxiliary respiratory musculature and marked stiffness and tenderness at the back of the neck, motor system with quadriparesis one/fifth (1/5) in superior, two/fifth (2/5) inferior extremities. Muscle reflexes were increased in arms and legs, bilateral positive Babinsky's sign, ankle and knee clonus and sensory hypoesthesia impairment, inferior to the first thoracic level were detected. The rectal sphincter tone was normal. Her family history revealed no unhealthy parents, only a 2-year-old sister with stroke-like events moya moyo disease with the onset at 1-year-old with ischaemic repetitive events.

INVESTIGATIONS

Laboratory studies including a complete blood count (CBC), serum electrolytes, blood urea nitrogen, creatine, urinalysis and glucose were all normal. Prothrombin time (PT)/activated partial thromboplastin time (apTT) was normal excluding

coagulopathy. The lumbar puncture revealed blood in the cerebrospinal fluid.

A sagittal T2-weighted MRI of cervical spine showed mixed signal intensity within spinal cord consistent in an intramedullary lesion of focal haemorrhage at levels C3–C6 and intensity changes below C6 vertebral segment (figure 1).

Evaluation of the spinal cord vessels using multislice CT angiography was performed 1 month later and disclosed a large anterior spinal aneurysm in levels C3–C6 with displacement of the spinal cord. The sagittal view showed a large aneurysm occupying almost the total width of the spinal canal at levels C3–C5 and extending down to medial margin of the sixth cervical vertebra. It was 30 mm length×18 mm wide×25 mm depth (figure 2). The feeding artery appeared to originate from the anterior spinal artery. Above the large aneurysm there is a small one specifically at level C2 of 0.7 mm length×0.5 mm wide, it was also supplied by the anterior spinal artery. The caudal portion of the aneurysm gives rise to two arterial sinusoidal branches (figure 3). A scan over the aorta segments resulted normal to excluding an aortic coarctation. The sagittal view with MIP (maximal intensity of projection) confirmed the presence of a large aneurysm after administration of contrast medium (figure 4).

DIFFERENTIAL DIAGNOSIS

A presumptive diagnosis of possible intramedullary arteriovenous malformation or hemangioblastoma and angioma was performed.

TREATMENT

Operation: a posterior microsurgical decompression C3–C6 laminectomy was performed and the dura mater was opened showing a reddish arachnoid. The spinal cord was opened at the posterior midline in dorsal median sulcus under an operating microscope 2 cm length and the findings of characteristic lesion of intramedullary arterial aneurysm was visualised, the wall was punctured using a fine needle resulting in reddish bleeding. Resection was considered inoperable at the time, therefore surgery was stopped, feeding vessels could not be identified and duroplasty was performed using muscle aponeurosis.

OUTCOME AND FOLLOW-UP

Postoperative course: the patient was admitted in the intensive care unit with mechanical ventilatory

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Figure 1 Sagittal T2-weighted MRI of cervical spine. (A and B) Mixed intensity within spinal cord at C3–C6 level consistent with intramedullary lesion of focal haemorrhage, (C) note the presence of a vascular structure below C6 vertebral segment.

support for 4 days. Respiratory and motor functions were improved progressively.

A further surgical approach was proposed and the parents refused it. The patient was monitored as an outpatient, over 1-year follow-up with physical therapy. She had no recurrence of clinical spinal subarachnoid haemorrhage and motor recover of superior extremities three-fifths (3/5) and inferior extremities three-fifths (3/5).

DISCUSSION

Spinal aneurysms in the high cervical region are extremely rare and are usually associated with spinal cord AV malformations

and coarctation of the aorta, both situations associated with increased blood flow. As an isolated entity, they are a rare occurrence. Few cases of isolated spinal aneurysms have been reported especially in children and the diagnosis can be a true challenge to the neurosurgeon.

The cause of spinal aneurysms is different to that of intracranial aneurysms. The primary mechanisms for development of spinal aneurysms are haemodynamic factors such as AV malformations with an increase of blood flow, and coarctation of the aorta where the anterior spinal artery is a significant route for collateral circulation.^{1–14} The mechanism for development of the isolated spinal aneurysms is not clear but it can be related to congenital vessel abnormalities of a genetic origin.⁸ The relation among this illness and that of her sister strongly suggest a common genetic origin.

One difference between spinal artery aneurysms and common intracranial aneurysms is that the former do not occur in bifurcation points.¹⁰

The pathology of spinal aneurysms has been poorly reported,^{10 12 15 16} but there has been report of the loss of elastic fibres and tunica media in the wall of the aneurysm. Kito⁷ reported a case associated with elastic pseudoxanthoma. Saunders¹² reported a case in association with fibromuscular hyperplasia, and others such as Goto¹⁷ considered a congenital abnormal arrangement of the vasculature.

The arterial supply to spinal aneurysms could come from several sources depending on each segment and localisation but there are few cases in the literature that allow the classification of spinal aneurysms in terms of vascular supply.

The clinical spectrum is wide and depends on the cranium–caudal locations of lesions; when it is found in the cranium–cervical junction, it often causes subarachnoid haemorrhage that resembles an intracranial ruptured aneurysm because the haemorrhage invades the cranial cavity.^{15 17–19} Spinal aneurysms in high cervical segments can be presented with headache, stiff neck, tenderness and manifestations of spinal cord compression.^{5 11 16 17 20 21} In the thoracic and lumbar regions the patients can report intense back pain and cranial symptoms.^{7 8 10 15 16 18–23}

Figure 2 Multislice CT angiography of cervical spine sagittal view showing a large spinal aneurysm occupying C3–C6 vertebral segments, note that lesion was almost totally occupying wide spinal canal.

Figure 3 Multislice CT angiography of cervical spine, coronal view (A–C) and axial (D) view showed: (A) spinal anterior artery feeding large and small aneurysm, note exactly localisation of small aneurysm in C2 vertebral segment; (B) presence of one of the two sinusoidal arterial branches in caudal segment of large aneurysm; (C) view of another arterial sinusoidal branches emerging from large aneurysm and (D) demonstrated small aneurysm and anterior spinal artery supplying both aneurysms.

The diagnosis can be demonstrated by a spinal angiography which is the preferred method.^{6 24 25} However, a multislice CT angiography is an excellent option to demonstrate this entity, especially when the selective spinal angiogram is not available. This case seems to be the first reported of a spinal aneurysm for this method.

Our case did show relative improvement after the decompressed effect of a laminectomy and duroplasty. This case also

seems to be very rare, to our knowledge, not only for being an isolated spinal aneurysm but such dimensions in a child.

The radical treatment of these aneurysms is naturally difficult, if not impossible, and the incidence and natural history are not well known, in contrast to their intracranial counterparts. There are, however, a few cases described with total resection. Nevertheless in our case there are several considerations, the endovascular occlusion of both anterior spinal aneurysms can

Figure 4 Multislice CT angiography MIP of cervical spine sagittal view showed large aneurysm after administration of contrast medium.

result in ischaemic spinal cord damage and other considerations for an anterior microvascular approach implies technical difficulties for possible vascular anastomosis and cervical bone stabilisation in a child of 1 year. The posterior approach by incision of dorsomedial sulcus can result in spinal cord damage by hematomyelia, retraction and ischaemia; therefore, we have awareness of high risk of cervical spinal cord damage and respect the parents' decisions for conservative management.

Microsurgery may be more efficacious in completely eliminating the aneurysm and its effects more durable over the extended lifetime of paediatric patients. Microsurgical therapy led to higher rates of complete obliteration and lower rates of recurrence, indicating an advantage over endovascular therapy in the critical areas of efficacy and durability. Furthermore, the incidence of delayed de novo aneurysm formation is considerably higher in patients treated endovascularly. Although the reason for this finding is unclear, it may relate to altered haemodynamics after endovascular interventions or direct vascular injury secondary to intraluminal microcatheter manipulation.

Parental biases should be addressed with serious discussions concerning the durability of treatment, delayed complications, angiographic surveillance and the potential for additional treatment later in life.^{14 25}

Definitive and durable treatment associated with microsurgery may outweigh the appeal of endovascular therapy when the extended life expectancy of these patients was considered.

Learning points

- Development of the sudden onset of quadriparesis state and neck pain should be considered as differential diagnosis of spinal arterial aneurysm in a child.
- A multislice CT angiography is an excellent and not invasive option to demonstrate this entity.

Contributors All the authors were involved in conception, design, acquisition of the data and analysis. They have reviewed critically the intellectual content and finally approved the last version to be published.

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