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CASE REPORT

Type IV Spinal Arteriovenous Malformation in a Child

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ABSTRACT

A 4-year-old boy presented with motor weakness and was subsequently found to have intraspinal extramedullary arteriovenous malformation (type IV C arteriovenous malformation) at the cervical region. Angiographic embolisation was performed. This patient illustrates the clinical inconspicuousness of spinal arteriovenous malformation in young patients and the importance of making the correct diagnosis so as to facilitate further interventional therapy.

Key Words: *Angiography, Arteriovenous malformation, Embolization, Intramedullary, Intraspinal, Magnetic resonance imaging*

CASE REPORT

A boy presented at the age of 4 years and 2 months with limb weakness and poor bowel control for 2 weeks. He had a history of asthma and had been given a 2-month course of steroids in The Philippines. The developmental milestones were unremarkable. At clinical examination, he was found to have a Cushingoid face with hirsutism. His gait was unsteady with a limp on the left side. He had weak limb power, affecting both the upper and lower limbs (power 4 minus/5) with normal tone, brisk ankle jerk, and upwards plantar reflex. The child was suspected to have iatrogenic Cushing disease, and short Synacthen test showed marked exogenous suppression. The electroencephalogram showed no abnormal signal, and computed tomography of the brain revealed no intracranial lesion. Magnetic resonance imaging (MRI) of the brain was also normal.

MRI of the cervical spine was then arranged for screening of an intraspinal lesion. It revealed a 2 x 2 x 4 cm intraspinal extramedullary vascular lesion, posterior in location, extending between C2 and C6 (Figure 1). This vascular lesion had slow flow, with significant extrinsic compression on the spinal cord. Contrast-enhanced

Figure 1. Magnetic resonance T1-weighted image — sagittal view.

MR angiogram confirmed that the vascular supply was coming from the anterior spinal arteries, which in turn were supplied by the left vertebral artery and the right intercostal artery (Figure 2).

Spinal angiogram was performed shortly afterwards and confirmed the diagnosis of intradural perimedullary

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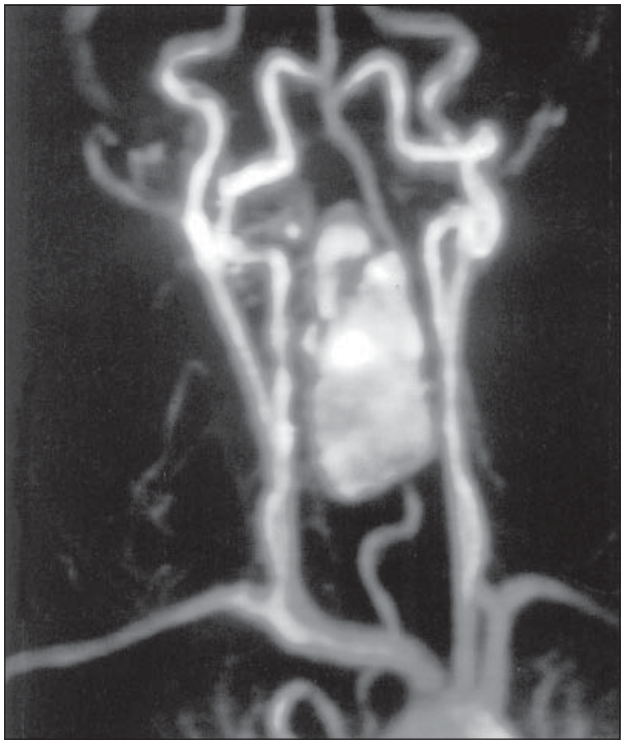

Figure 2. Contrast-enhanced magnetic resonance angiogram — frontal view.

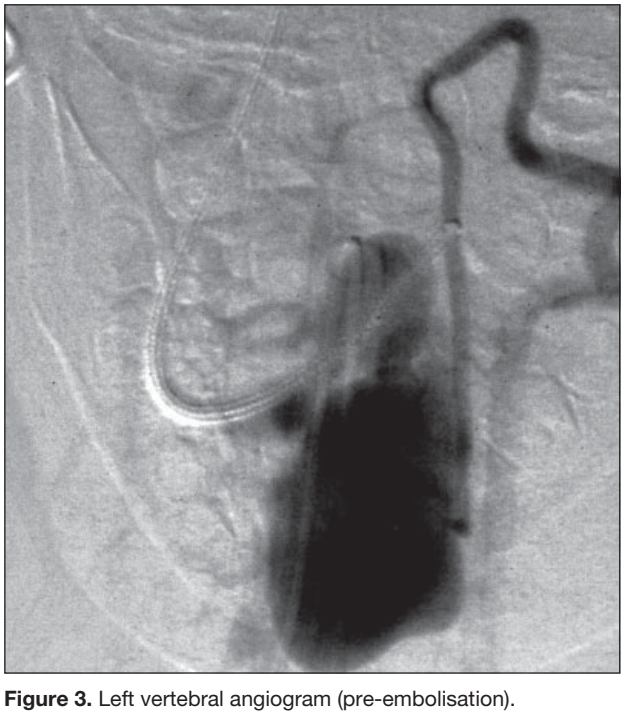

Figure 3. Left vertebral angiogram (pre-embolisation).

direct arteriovenous (AV) fistula (type IV AV malformation). A single hole AV fistula was noted in the spinal canal at the cervical region, which led to a huge venous pouch occupying the enlarged bony spinal canal. The fistula received a blood supply from 2 arterial feeders directly via the anterior spinal arteries

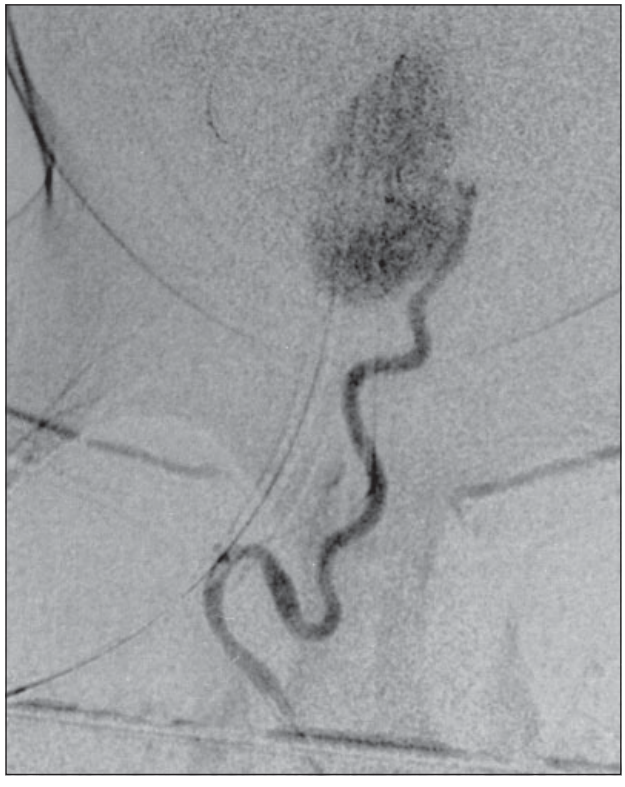

Figure 4. Right subclavian angiogram (pre-embolisation).

arising superiorly from the left vertebral artery and inferiorly from the right upper intercostal artery (Figures 3 and 4). The venous pouch drained into the perimedullary venous plexus via a tortuous tributary.

Embolisation of the fistula was performed. Firstly, systemic heparinisation was given and a 4F vertebral Optitorque catheter was used as a guiding catheter. The upper feeder from the left vertebral artery was catheterised with Excel-14 aided by Transend-14 GW and was embolised with a 2 x 4 mm Vortx GDC-18 followed by glue (50% histoacryl mixed with lipiodol). The second feeder from the right upper intercostal artery was embolised with a 3 mm x 12 cm GDC-10 followed by glue. Postembolisation angiogram showed that the dual arterial feeders were completely occluded and there was no filling of the venous pouch (Figure 5).

MRI was repeated for postembolisation follow up 6 weeks later. The contrast-enhanced MR angiogram showed that the AV fistula was diminished in size and contained old blood products. Contrast-enhanced MR angiogram found no recanalisation of the arterial feeders. Clinically, the limb power improved after the embolisation to grade 5 minus/5. The tendon reflex was still brisk, and the patient could stand on 1 leg but was unable to hop.

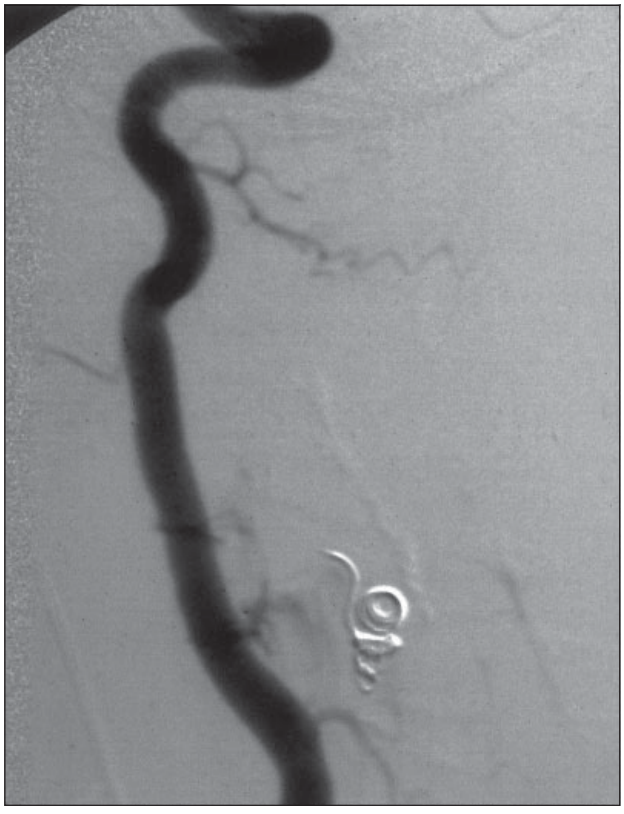

Figure 5. Left vertebral angiogram—frontal view (postembolisation).

DISCUSSION

Spinal vascular or AV malformation (AVM) is classified into 4 types.[1] Type IV AVM was first described by Djindjian et al[2] and represents approximately 10% to 20% of all spinal AVMs.[3] Type IV AVM represents an intradural, extramedullary, or perimedullary direct fistula without nidus. It is predominantly found in the thoracolumbar region,[4] and the lesion is usually located on the ventral surface of the cord. The arterial supply usually comes from the anterior spinal arteries, and blood supply from the posterior spinal artery is less common. Three subclasses (A, B, and C) have been renamed by Anson and Spetzler,[5] and the subclassification depends on the size of the arterial feeders. Subtype A is characterised by a normal-sized spinal artery with slow flow and mild venous engorgement. A subtype C lesion comprises a large fistula with dilated high-flow arteries and a tortuous draining vein. Subtype B is considered to be an intermediate lesion between subtypes A and C. Subtype A may cause venous hypertension due to the small fistula and slow flow. Subtypes B and C may lead to arterial steal, and spinal compression due to the involvement of large vessels. Subarachnoid haemorrhage has been reported in 10% to 20% of type IV AVMs, and usually occurs in conjunction with subtypes B and C.[4,6]

Flow void signals are present at MRI and are sometimes prominent, causing confusion with other types of AVM.[7] The distended vessels may cause significant compression on the spinal cord. However, if the lesion is too small to be visualised by MRI, myelography should be done for better visualisation.[7,8] MR angiography is an effective method of diagnosing spinal AVM and to image arterial feeders, nidus, and venous drainage.

Angiography is mandatory before endovascular intervention, and it is necessary to perform thorough spinal angiography to delineate both the spinal feeder and the dural branch feeder.[9] As shown in this patient, there were 2 arterial feeders from the left vertebral artery as well as the right subclavian artery and the contrast-enhanced MR angiogram provided valuable guidance for the latter angiogram, in which the feeder from the right subclavian artery was shown.

Endovascular embolisation is a useful adjunct to surgery for large and high-flow AVMs,[10] but different subtypes require different treatments. Type A arterial feeders are usually of normal calibre, and embolisation is difficult, leaving surgery as the only treatment option. Type B and C subtypes are usually accessible for endovascular embolisation.

Contrast-enhanced MR angiography is more sensitive than MRI for depicting residual or recurrent flow in perimedullary vessels and indicates the patency of the vascular malformation.[11] MR angiography is acceptable for imaging follow-up for clinicians and patients.

Type IV AVMs are thought to be congenital. Familial occurrence in 2 young siblings complicated by pseudoaneurysm of the thoracic cord has been reported.[12] Some cases are associated with trauma.[13-15] Spinal AVMs have also been reported to be associated with Rendu-Osler-Weber syndrome,[7] and it could be the first manifestation in a child with this disorder.[16]

The age of presentation ranges from 1 to 70 years,[6,17,18] and the clinical symptoms are usually progressive ascending myelopathy, with sphincter dysfunction. If slowly progressive or acute radicular or medullary symptoms arise in children, spinal AVM should be ruled out by MRI.[19] Perimedullary fistulas mostly deteriorate quickly because of haemodynamic or haemorrhagic problems.[20]

As for this child who was initially diagnosed with iatrogenic Cushing disease causing neuropathy and limb

weakness, the diagnosis is only clear after MRI. This case, subclassified as a type IV C spinal perimedullary AVM, was uncommon given its location at the cervical region and the patient's age at presentation. Another similar condition, which was initially misdiagnosed to be Guillain-Barre syndrome, has been reported.[21]

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