

CASE REPORT

Spinal dural arteriovenous fistula in a case with lipomyelodysplasia

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ABSTRACT. Coexistence of a spinal dural arteriovenous fistula within a dysraphic spinal lesion is a very rare situation. We report a 40-year-old man who presented with low back pain and progressive paraparesis. MR images showed an intradural high signal intensity mass at the L2–L3 level containing irregular signal void structures. Spinal angiography revealed extradural arteriovenous fistula with three connections, drained by a tortuous perimedullary vein.

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Spinal vascular malformations (SVM) are rare diseases compared with vascular malformations of the brain. SVM can be differentiated into congenital lesions (spinal cord arteriovenous malformations (AVMs) and cavernomas) and acquired lesions (dural arteriovenous fistulae (AVFs)) [1]. Association of SVM with a dysraphic spinal lesion is extremely rare which makes the management more complicated [2, 3]. Embolic occlusion is the preferred method for most of the SVMs, achieved using particulates, such as polyvinyl alcohol particles, or liquids, such as isobutyl 2-cyanoacrylate [4]. We report a case of a combined spinal arteriovenous malformation and lipomyelomeningocele, emphasising the diagnostic features and embolisation procedure performed with ethyl vinyl alcohol co-polymer (Onyx).

Case report

A 40-year-old man presented with a 1 year history of lower back pain and progressive fatigue in both legs precipitated by physical exertion. He was also complaining of a number of ulcers approximately 2–4 cm in diameter, over the knee and anterior surface of the crural and ankle region of both legs sparing the feet. Ulcers were suggestive of a repetitive thermal injury. On physical examination, there was a brown-red discoloration on the patient's back. Neurological examination showed loss of pain and temperature sensation over the L4 and L5 dermatomes accompanied by bilateral leg weakness. MRI of the thoracic and lumbar spine was performed. MR studies revealed on T_1 and T_2 weighted images an intradural high signal intensity mass containing irregular signal void structures representing vascular

channels in a lipoma (Figure 1). The lipoma containing vascular and cord elements was tethering the spinal cord and extending dorsally through a dysraphic cleft at L3. Terminal hydromyelia was also noted extending over three segments. Dilated perimedullary vessels were seen extending along the surface of the spinal cord up to the mid-thoracic level. Spinal angiography showed the fistula fed by the second right lumbar artery. A small calibre feeder was also noted arising from the left third lumbar artery. The delayed phase showed a tortuous perimedullary drainage vein. With these clinical and radiological findings, before the surgical intervention directed towards the lipoma and tethered cord, we decided to embolise the fistula first.

Embolisation

With the patient under conscious sedation and systemic anti-coagulation, embolisation was performed through the right L2 and left L3 lumbar arteries. First, a diagnostic 6 F Simmons 1 catheter was placed at the proximal segment of the lumbar artery. A dimethylsulphoxide (DMSO)-compatible microcatheter (Ultraflow; MTI Corp., USA) was then advanced coaxially into the radiculodural feeders under road-mapping with a microwire (Mirage; MTI Corp., USA). The dead space of the microcatheter was filled with DMSO over 1 min, then, under real-time road mapping, 0.8–1 ml of 6% ethyl vinyl alcohol co-polymer (Onyx; MTI) was injected into the sinus slowly, by a "reflux-and-push technique" as used in the intranidal Onyx injections for brain arteriovenous malformations [5]. This procedure was repeated for each feeder. The injections stopped as soon as the Onyx reached the proximal intradural draining vein of the fistulae. A total of 2.6 ml of Onyx was used. The first injection performed from the left side was less than 10 min and the second injection from the right lumbar artery was just under 20 min. We obtained several arteriograms through the

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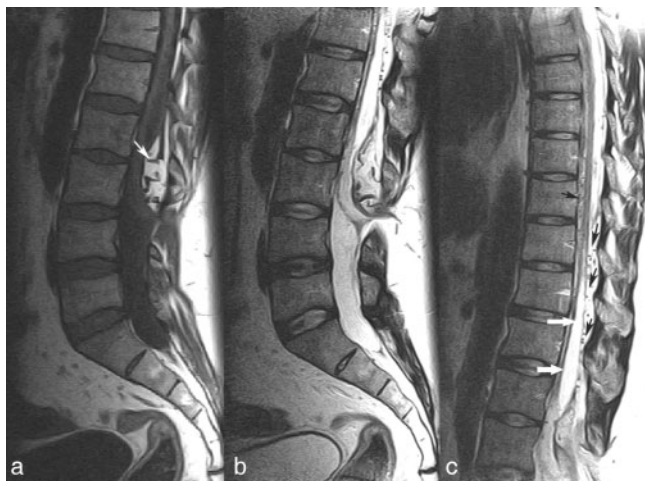


Figure 1. MR images of lumbar and thoracic spine. Sagittal (a) T_1 weighted and (b) T_2 weighted images show multiple signal voids within an intradural lipoma (arrow). (c) Sagittal T_2 weighted image shows the lipoma tethering the cord with terminal syrinx (white arrows) and the dilated perimedullary vessels (black arrows).

catheter to ensure obliteration of the fistula. On demonstration of closure of the fistulae, the microcatheter was straightened and pulled out rapidly with a little movement of the Onyx cast. Angiograms were repeated after microcatheter withdrawal from the lumbar artery to confirm the total obliteration of the fistulae. Heparinization was not continued after the embolisation. The patient did not develop any new neurological deficits. 1 month after the embolisation procedure the patient underwent surgery. Following L2–L3 laminectomy the subcutaneous fat tissue was removed until its entrance into the thecal sac. A dorsal vertical durotomy was performed which revealed the intramedullary portion of the lipoma that was harbouring embolised tangles of vessels. A subtotal resection could be performed because intramedullary lipoma was tightly adhered to surrounding structures. The lesion was untethered and the dural

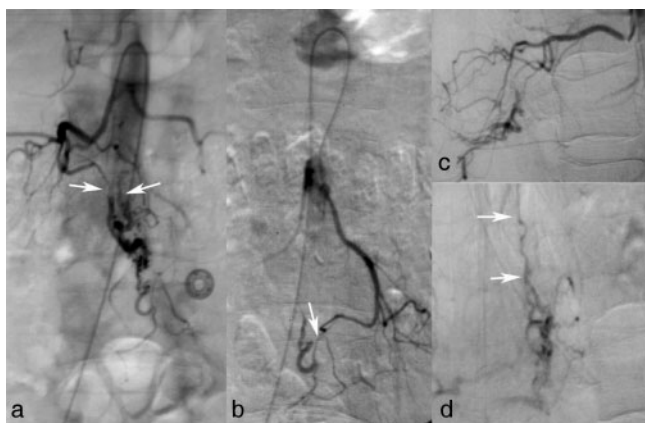


Figure 2. Spinal angiograms. (a) Anteroposterior angiograms obtained from the right L2 artery show the two fine feeders and the fistula sites (arrows). (b) Left L3 injection shows one more fistula. (c) Lateral angiogram of the right L2 injection shows the extracanalicular location of the fistula. (d) Sagittal late phase image shows tangles of veins forming a dilated tortuous perimedullary vein (arrows).



Figure 3. Angiograms and spot images obtained during embolisation. (a) Anteroposterior view obtained from the right L2 lumbar artery injection after the embolisation from left L3 shows the remaining fistulae. (b) Anteroposterior spot image shows the Onyx cast of the left L3 lumbar artery embolisation. Note the fine filling of the fistula with a considerable amount of venous reflux. (c) Angiogram from the right lumbar artery shows the elimination of the fistula and the final Onyx cast.

reconstruction was accomplished. 2 months after discharge he was free of his back pain and showed complete recovery of his leg weakness.

Discussion

Spinal lipomas with spinal dysraphism (intraspinous lipoma, lipomyelocele or lipomyelomeningocele) result from focal premature separation of epidermal ectoderm from neural ectoderm (neural tube) which further facilitates incorporation of mesenchyme between the closing neural tube and skin. This mesenchyme could prevent closure of the neural tube focally. Moreover, the dorsal surface of the closing plate induces differentiation of the mesenchyme primarily into fat. Because of the pluripotent nature of the mesenchyme a number of other tissues can be noted within the substance of lipomyelomeningocele including striated muscle, cartilage and fibrous bands. However, vascular malformations are unexpected in this setting [6].

Only four cases of combined spinal lipoma and spinal vascular malformation have been reported. Two of them were intramedullary AVMs embedded in intradural intramedullary lipoma [3, 7]. The other two were dural AVFs, one of which was combined with sacral filum terminale lipoma [8] whereas the other AVF was within a lumbar lipomyelomeningocele located extraspinaly [2]. Spinal AVMs are congenital lesions including intramedullary (*i.e.* glomerular and juvenile) and perimedullary (fistulous) AVMs. On the other hand late and gradual presentation of symptoms and lack of association with other vascular or non-vascular malformations are main features of spinal dural AVFs suggesting an acquired process [1]. In our case besides clinical presentation, features such as extracanalicular location of the shunts, lack of a nidus formation are suggestive of a dural AVF rather than an AVM.

The majority of patients with lipomyelomeningocele are neurologically intact at birth, but progressive

neurological deterioration develops at younger ages. Late presentation as in our case is very rare [9]. However, the two previous cases with combined spinal lipomyelomeningocele and spinal vascular malformation also presented late in life [2, 3]. Severity of the tethering process seems to be closely related to the magnitude and presentation of the symptoms. Lower back pain and progressive motor weakness were the main neurological problems of our patient. These symptoms were presumably related to tethering of the cord and the caudal elements by the growing lipoma mass with time. Symptoms may also be attributed to venous hypertension as a consequence of AVF or comorbidity of the terminal syrinx as in our patient. Time lag between the embolisation and surgical procedure showed that the tethering was the main pathophysiology.

Surgical obliteration or excision of spinal dural arteriovenous fistulae has a high rate of clinical success with an acceptable morbidity. However, embolisation when feasible also has favourable outcomes and besides offers less invasiveness and earlier rehabilitation [4, 10]. In the present case surgical exploration and removal of the fistulae embedded in the lipoid mass was quite difficult. Embolisation made the dissection and subtotal resection of the mass much easier.

In conclusion, although very rare, lipomyelomeningoceles can be associated with spinal vascular malformations. Embolisation of the vascular malformation would facilitate the surgical dissection of the mass.

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