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Intradural spinal arteriovenous malformation of the glomus type: a case report

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Summary

Spinal arteriovenous malformations (AVMs) being even rarer than cranial ones, constitute 3 – 4% of all spinal tumours, with an overall male:female ratio of 4:1. We report the case of a 24-year old female Youth Corps member who presented with an apoplectic onset of a left hemiparesis, progressively deteriorating to an incomplete quadriplegia with motor power of 2 and 0 in the right and left lower limb muscle groups, respectively. Magnetic resonance imaging (MRI) showed a vascular malformation whose precise nature was indeterminate; the definitive diagnosis of a Glomus AVM was only made intra-operatively. Laminectomy with complete surgical excision was done and she gradually improved to ambulate with minimal support. The experience in our unit tends to support the observation in literature that spinal arteriovenous malformations could be very rare, and depending on the location, could be amenable to complete surgical excision and recovery of neurological function.

Keywords : *Glomus arteriovenous malformation, spinal cord, angiography, laminectomy.*

Résumé

Les malformations de l'arterioveineux Vertebral (MAU) etant meme plus rares que les cranienne constituent 3-4% de toutes les tumeurs vertebrales avec une proportion male: femelle totale 4:1 de toutes les tumeurs vvterbrales. Il y' a le cas d'une fille de 24ans member du corps des volontaires qui qvait un debut apolplectique d'une hemiparesu gauche et qui se deteriorait progressivement a une quadriplegie incomplete avec les muscles droit et gauche du member inferieur des groupes. L'image de la resonance magnetique (IRM) a montre une malformation vasculaire don't la nature precise etait indeterminee, le diagnostique definitif d'un glomus MAV a ete fait intra-operatoirement. La laminectomie de copure chirugicale, complete a ete fait et elle s'est amelioree progressivement par le support ambulatoire minimal. L'experience dans notre unite a tendance a supporter l'observation dans la literature et montre que les malformations de l'arterioveineux vertebrales pourrait etre rares et selon l'emplacement, pourrait etre responsible pour completer la conpure chirugicale et la recuperation de la fonction neurologique

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Introduction

Spinal arteriovenous malformations (AVMs) are rare, and lack pathognomonic features. They are known to mimic lesions ranging from those causing raised intracranial pressure to those of acute spinal cord trauma [1,2] and purely clinical diagnosis is rare.

Spinal tumours occur in 3 to 10 per 100,000 population, which is a fifth of that of brain tumours [3]. The age range at diagnosis is 11days to 74 years [4] with nearly 80% presenting between 20 – 60years of age; 70% of the lesions are located in the cervical and upper thoracic spinal cord with almost an equal male:female ratio [5,6].

Of these, about 60% are intradural and mostly, extramedullary (80%) in adults. The paediatric occurrence of extra- and intra-medullary tumours is equal [7]. Ninety per cent of all intradural tumours are benign and potentially respectable [8].

Spinal AVMs constitute about 3 - 4% of intraspinal masses and 10% of the incidence of cerebral AVMs [9]. They are classified into: Type I – Dural Arteriovenous Fistula, DAVF; II – Juvenile AVM; III – Glomus AVM, and IV – Direct Arteriovenous Fistula, AVF [10]. Overall, the male:female ratio is 4:1 [11] though this varies with the type of AVM.

Dural AVF (Type I): The lesion in this type is restricted to the dura mater, with the fistula embedded in the dural covering of the nerve root and the adjacent spinal dura. Ninety per cent of the patients are males above 40years, presenting with paraparesis and bisphincteric dysfunction, of gradual onset [2]. On account of these characteristics, a DAVF is thought to be acquired in origin [12] unlike the intradural type which is regarded as congenital.

Intradural AVM (Types II – IV): These lack sex predilection, present below 40years with apoplexy and tend to occur more in the cervical and upper thoracic cord. Forty-four per cent are associated with aneurysms [13]. The nidus in this type is either within the tissue of the spinal cord or is embedded in the pia. These are further classified into: Juvenile (II) and Glomus (III) - both of which constitute 15 - 20%, and Direct AVFs (IV) – 80% [14].

The Juvenile (Type II) is supplied by any of the medullary arteries, has a large nidus and as a rule, contains cord tissue interspersed among the AVM vessels, and sometimes involves the vertebral and paraspinous soft tissue, with an audible bruit at the point of maximum shunting.

The Glomus (Type III) has a nidus of tightly packed mass of blood vessels confined to a short segment of the cord. They occur mostly in the anterior spinal cord, making surgical access difficult.

Direct AVF (Type IV) are high-flow extramedullary lesions with a direct shunt from an artery to a vein, and are located on the surface of the cord or free in the subarachnoid space [15]. They are associated with large venous or arterial aneurysms compressing the cord or causing symptoms by vascular steal, haemorrhage or venous congestion, producing various cord syndromes. Direct AVFs are further divided into small, larger and giant fistulae, thus graded as IVa, IVb and IVc, respectively.

The case we are reporting is an intradural spinal AVM of the classic Glomus type (III), managed operatively in our Unit in 2004.

Case report

Our patient is a 24-year old female fresh graduate who was referred to us in April 2004 from our Neurology Unit. She had presented to the Neurologists with a 3-day inability to use her left upper and lower limbs. The history was that of a sharp pain radiating from her left upper limbs down to her left foot, woke her up from sleep, only to discover that she could not use her left limbs. She also had urinary urgency and constipation. She had experienced neither headache, nor abnormalities in her level of consciousness or mental functions. Other systems were normal.

She had experienced neck pains and stiffness on and off in the 6 years before presentation, but these had been generally mild and resolved early. She was not on any regular medications. She was single and neither used alcohol nor tobacco. Her psychiatric history revealed no gross abnormalities.

Physical examination (2 months since event) revealed an anxious lady with no general features of chronic systemic ailments. Her nervous system was normal except for the long tracts which revealed mild muscle wasting on all limbs and the trunk, worse on the left half. There were no abnormal spontaneous movements. Limbs were hypertonic and paretic; power on the right half ranged from 3 – 4, and on the left 0 – 2 (worse in the lower extremities); cerebellar functions were normal, muscle stretch reflexes were hyperreflexic with extensor response to plantar reflexes. Sphincters were normal. Sensory level was C3 / C4 with residual to S5. Skull, spine as well as other systems were normal.

The initial diagnosis was that of cervical cord compression from a space occupying lesion (?neoplastic ??granulomatous).

Urinalysis, haemogram and blood chemistry were normal. Helical computerized tomographic (CT) myelography of the cervical spine showed an expanded segment of the spinal cord with serpenginous intra-/extra-medullary lesions between C3 - 6 levels. This led to the consideration of (unusual) vascular neoplasm, AVM or parasitic infestation.

Magnetic resonance imaging (MRI) did not give substantial additional information, though it showed the serpenginous lesions more clearly with its left-sided preponderance (Fig 1).

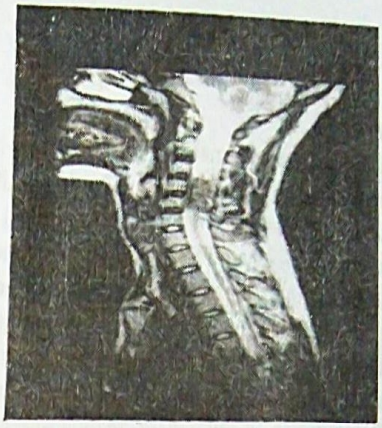

Fig. 1:

The patient was worked up and cervical laminectomy done.

At operation, laminectomy of the third, fourth, fifth and sixth cervical spines was done, and the findings were - huge coils of extramedullary vascular dilatations, mostly arterialized coronal veins, with intramedullary extension (Fig. 3). There was associated resolving subarachnoid haemorrhage. Unprepared for an extensive vascular procedure, and considering the late time of day we applied a fascial dural patch and closed up after a further evaluation of the lesion, with a view to doing angiography.

The patient's neurological status, however, continued to deteriorate over the weeks with motor power globally worse (0 - 3) while we eventually did a helical CT subtraction angiography.

The helical angiography identified the spinal intradural AVM with a prominent feeding branch from the left posterior spinal artery (Fig. 2). It did not reveal an aneurysmal dilatation.

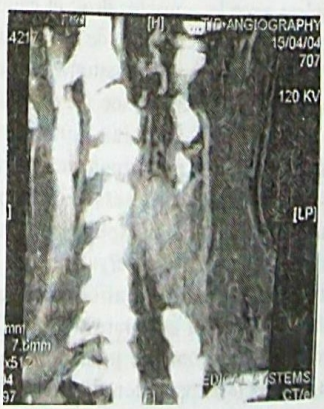

Fig. 2:

At re-operation 5weeks later, laminectomy was rostrally extended to C2 and caudally to C7. A large radicular artery with multi-level medullary arteries was found feed-

ing the lesion, with an intramedullary glomus of tightly packed vessels (without interspersing cord tissue at C5 – 6 cord level (Fig. 3).

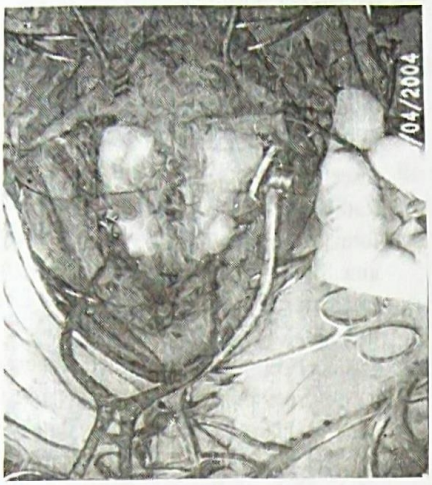

Fig. 3

There was no associated aneurysm. A dorsal longitudinal myelotomy, with bipolar coagulation and ligation of the feeding radicular and medullary arteries, was done. A gross total excision of the glomus and nidus was done with the aid of operating loupes, and the specimen was sent for histological studies. In the immediate post-operative period, the patient had a complete left lung collapse, which resolved with fibreoptic bronchoscopy, suctioning and chest physiotherapy. She was ambulated on a wheelchair with the aid of a rigid neck collar on the 5th post-operative day, and made progressive but slow neurological gains. At discharge from hospital 5weeks post-operation, power on the right was 4 - 5^- and on the left 2 - 3^+. In the clinic, 6weeks after discharge, motor power in all the limb muscle groups had improved to 4 – 4^+, and by 3months post-discharge power was 5^- - 5 globally; she had begun ambulating with the minimal support of a walking stick. Post-operative angiography could not be done due to lack of funds. Histology report, however, confirmed an AVM.

Discussion

The rarity of spinal AVM in our practice appears to be global. The same is also true of the low suspicion of this lesion, at presentation. The usual demographic pattern is also well illustrated by our patient.

Because AVMs present as mass lesions, they are easily mistaken for other tumours that are common in different environments. Also, since they may present with haemorrhage, they could be confused with infective processes if subarachnoid spill has occurred, especially in our environment. Spinal subdural haematoma and peripheral siderosis have, less commonly, complicated AVMs becoming in some cases the major clinical presentation [13,14].

The tightly packed mass of coiled vessels without intervening cord tissue in the intramedullary nidus and the huge extramedullary arterialized coronal veins in our 24-year old female patient, all added up to a diagnosis of Glomus intramedullary (Type III) AVM.

In 40% of patients with IAVM, an acute presentation is caused by subarachnoid or intramedullary haemorrhage, while 50% would have had a subarachnoid bleed at the time of presentation [2] - a phenomenon commonly known as *Coup de Poignard of Michon*. Such were the clinical and operative findings in this case.

The history of recurrent neck pains with occasional stiffness for more than 6years, prior to the sudden onset of myelo-compression, was suggestive of a long existing lesion, with a tendency to bleed intermittently. The site of the bleed could, however, not be identified.

In the presented case, appropriate initial imaging was suggestive. A higher level of suspicion would have led to pre-operative angiographic studies especially with CT angiography which is technically easier and less hazardous than cannulation techniques. We agree, however, that selective arteriography will sometimes be inevitable.

Delay in diagnosis due to associated diagnostic difficulties has been recognized by Ratliff and Connolly [6], as one of the pitfalls in the management of AVMs.

Oldfield EH, in discussing spinal AVMs, advises that a complete diagnostic investigation of the spinal cord and precise diagnosis of the type of malformation is essential before a decision to undertake a surgical or embolic treatment because the different types of vascular malformations require distinct management protocols, and successful treatment of the malformation demands a pre-operative knowledge of the anatomy of, and the blood supply to the lesion and the spinal cord [2]. The paucity of information from both CT and MRI as in this case must have informed the opinion of Oldfield that the diagnostic approach should be performed in two stages : a) screening studies with MRI or myelography to detect or raise the suspicion of a vascular abnormality and, b) selective arteriography to confirm the diagnosis, precisely localize the lesion and define the vascular anatomy of the malformation and the blood supply of the spinal cord [2]. Identifying the cord blood supply is very necessary, to obviate the catastrophic neurological deterioration that could accompany the surgical interruption of a sole radicular or segmental arterial spinal cord supply.

Complete obliteration of the AVM should be the goal of all modes of treatment [2]. This was facilitated in our case by the posterior location of the nidus, with arterial supply from the left posterior spinal artery. Anteriorly situated lesions are not readily amenable to complete surgical extirpation because of the difficult access and likely complications. The alternative therapy is embolization which however, has a long term chance of re-cannalization even as early as 6months post-therapy [15]. Surgical excision seems, therefore, to be the preferred modality where this is feasible. Recently, however, Hamada and his co-

workers in Japan reported the successful use of cellulose porous beads as an embolic material for permanent vascular occlusion [16]. Also, Kuga, *et al*, reported the successful treatment, by cyanocrylate embolization, of a giant spinal AVM with multi-segmental feeding vessels from the 5th to the 12th intercostal arteries causing cardiac failure and thoracic myelopathy, with complete clinical and neurological recovery [17]. These developments, if they stand the test of time, would favourably influence the choice of embolic procedures over the more invasive and extensive routine surgical options, in the future.

Outcome of AVM treatment depends on the type and location of the lesion and the pre-operative level of neurological dysfunction. Oldfield stated : *Patients who are ambulatory before treatment are usually ambulatory after treatment.* This statement, has been corroborated by Biondi and several other workers [18,19,20]. Morgan MK, in assessing the outcome of treatment of the various types of AVM in a review of several series, concluded that : Type I responds well if surgical disconnection is done early; Type II is best treated by surgery with risks of a poor outcome; Type III is rarely treatable; and Types IVa/IVb are likely to respond well to surgical treatment, while IVc would likely fare poorly with surgery and so, may be treated by endovascular therapy [21]. These recommendations await future criticisms based on experience. We would wish to add that posteriorly situated Type III lesions would likely respond well to surgical excision, and that a patient who could not ambulate before treatment might still fully ambulate after treatment.

Post-operative arteriography is used to assess the success of complete excision. Post-embolization, Oldfield prescribes repeat arteriography 7 - 10days later [2] with follow-up studies every 1 - 2years, or when symptoms recur. We hope to do a follow-up angiography within this period.

Conclusion

This case underscores the ease with which a spinal AVM may be missed clinically, and so, the need to maintain a high index of suspicion, in order to avoid ignoring salient diagnostic pointers. It also underscores the importance of location and access, (apart from the type) of this uncommon lesion, in prognostication. Lastly, the case suggests that in spite of the challenging nature of definitive treatment, outcome can be rewarding even when patients are no longer ambulatory.

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