ACUTE PARAPARESIS IN A PATIENT WITH JUVENILE TYPE SPINAL ARTERIOVENOUS MALFORMATION

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ABSTRACT :

A patient presenting with sudden onset of backache, paraparesis and urine retention was found to have low thoracic intramedullary lesion on MRI. A juvenile-type arteriovenous malformation was observed through a posterior approach, but feeding arteries could not be found in spite of lateral and also anterolateral exploration. Multiple feeders via anterior and posterolateral spinal arteries were observed on the first spinal selective angiogram. Embolization could not be performed because of iatrogenic spinal subarachnoid haemorrhage. Embolization was achieved through a sulcal artery via anterior spinal axis during the second session. Patient's symptoms relieved progressively after the embolization. This result suggests that embolization is the preferred modality for treatment of the Juvenile–type spinal arteriovenous malformations (AVM's).

Key words: *angiography, embolization, spinal arteriovenous malformation, spinal cord.*

Running Title: *Juvenile type spinal arteriovenous malformation.*

INTRODUCTION

Spinal AVM's are heterogenous group of nonneoplastic vascular abnormalities which may derive from embryological errors. They cause acute, subacute or chronic medullary dysfunction which is either complete or partial. Spinal AVM's are seen in younger patients (less than 30 years). About half of the patients with spinal intradural AVM's have acute presentation caused by subarachnoid, intramedullary or epidural haemorrhage and the other half have a gradual onset and progression of symptoms and signs. The latter form of presentation results from watershed infarction due to arterial ischemia by a vascular steal phenomenon or a result of venous congestion. Other rare reasons of myelopathy are spontaneous thrombosis resulting with necrotizing myelopathy of Foix–Alajouanine disease and mass effect. Spinal cord AVM's can be classified into three types; dural arteriovenous fistulas (AVF's= formerly known as Type I or single coiled vessel), intradural AVM's and cavernous angioma. Intradural AVM's have been divided into three subcategories as juvenile AVM's (formerly Type II), and intradural (perimedullary) AVF's (formerly Type IV) (1, 4, 8, 9, 11).

In this study, a patient with juvenile–type AVM with a rapid onset of incomplete paraparesis is presented. The purpose of this study is to reveal the superiority of embolization among the treatment modalities of the spinal AVM's.

MATERIAL and METHODS

A ten years old girl was admitted to the hospital with a sudden onset of backache radiating to the lower extremities. In neurological examination, paraparesis (2/5, loss of sensation below L1 and urine retention were detected. ASIA Impairment Scale was used in grading the degree of impairment and found as C (15). Evaluation and monitoring of daily–life activities were achieved by functional independence measure (FIM) for determination of functional assessment of the present stage and follow–up. Total FIM was found as 66 at admittance. An enhanced intramedullary lesion at the lower thoracic region was demonstrated on MRI. There occurred a local swelling, caused by a intramedullary mass that appeared hyperintense on both T1 and T2–weighted images corresponding to a subacute haematoma (Fig. 1A, B) suggesting either a

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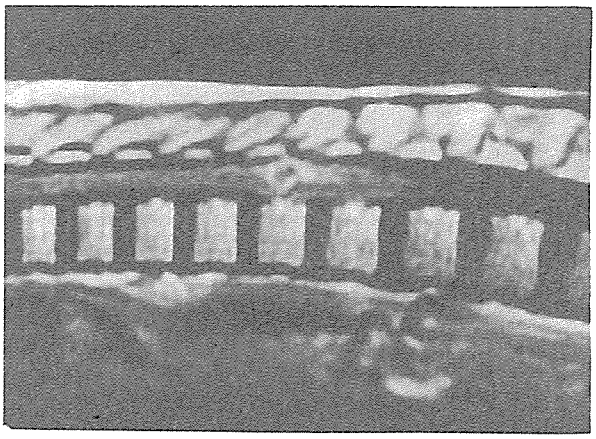

Fig. 1 A. Midsagittal T1–weighted MR shows lower dorsal intramedullary hyperintense lesion. There exists a local swelling.

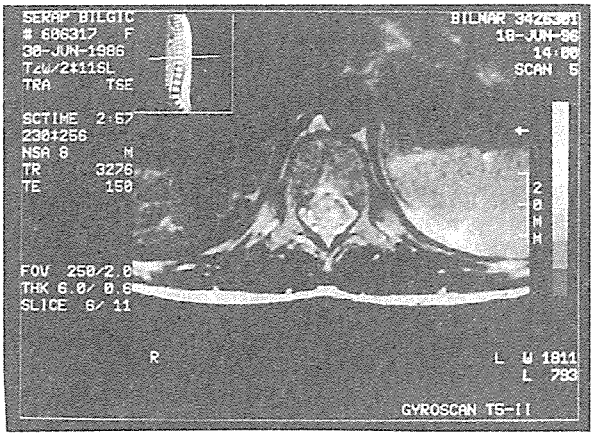

Fig. 1 B. Same lesion appears hyperintense on T2–weighted axial cut.

vascular malformation or an already bled tumor such as spinal ependymoma or haemangioblastoma. Patient was operated on immediately with osteoplastic laminoplasty because of the rapidly progression of paraparesis. A juvenile–type AVM was observed during the exploration process but no feeding arteries either dorsally or laterally were identified. No discoloration of the cord attributable to intramedullary or subarachnoid haemorrhage was observed. Paraparesis was slightly progressed (1/5) early after operation, but improved to same status on 5th postoperative day. Selective spinal angiogram was performed 12 days after the operation that confirmed an intramedullary AVM (Fig. 2). The AVM's blood supplies were mainly from anterior spinal artery and secondarily from posterolateral spinal artery. An intranidal aneurysm probably of venous origin was observed. That part of the nidus was accepted as the weak point of predisposing rehaemorrhage. Therefore, it was first thought to be treated by embolization but could not be performed because of the iatrogenic rupture of the anterior spinal feeder following the insertion of the catheter. Anyhow, no neurologic deterioration was observed after this iatrogenic subarachnoid haemorrhage. Second angiogram was performed a month later. A 10 Jetstream (Medtronic–CA, USA) microcatheter was positioned in the feeding sulcal artery via anterior spinal axis. A control angiogram confirmed the position of the catheter tip (Fig. 3). Hystoacryl Bleu 25% (Braun–Germany) and Lipiodol (Guerbet–France) mixture was injected resulting in total closure of that part of the nidus and the aneurysm. Control angiogram a year later showed occlusion of the aneurysm and intramedullary part of AVM (Fig. 4).

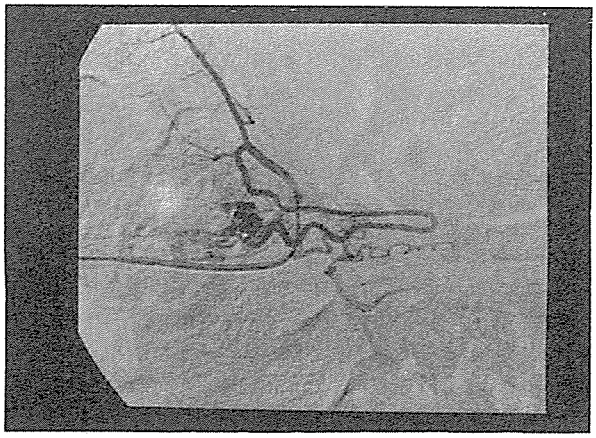

Fig. 2. Selective injection of the right D11 intercostal artery. AP projection shows an intramedullary AVM fed by the anterior spinal artery. Note the intranidal aneurysm and enlarged anterior spinal axis.

RESULTS

Patient's symptoms relieved progressively after the embolization. The patient recovered totally from her sensory deficit and was able to walk independently after consecutive physiotherapy sessions lasting a year (ASIA Impairment Scale=D, total FIM score=115). Nevertheless, she has automatic urinary bladder and has not regained sphincter control till the time being.

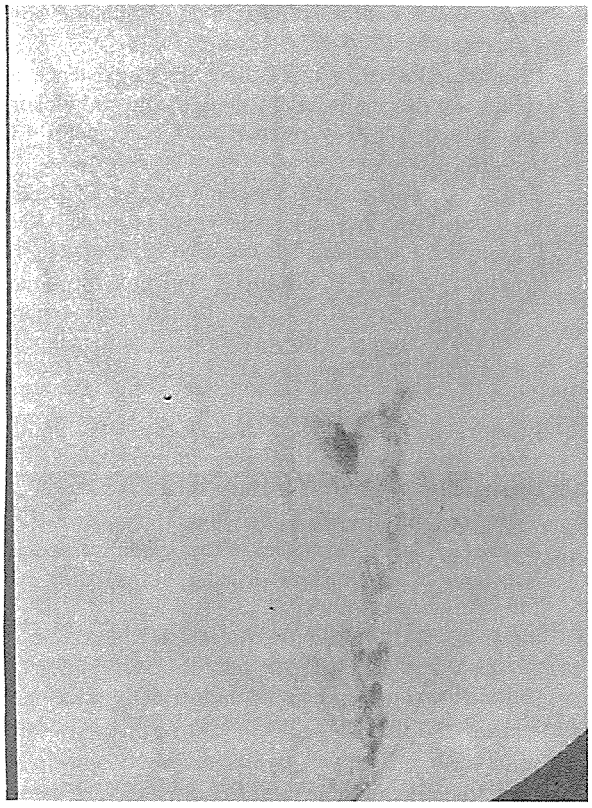

Fig. 3. Selective injection of sulcal artery feeding the malformation shows the nidus and multidirectional venous drainage either anteriorly or posteriorly.

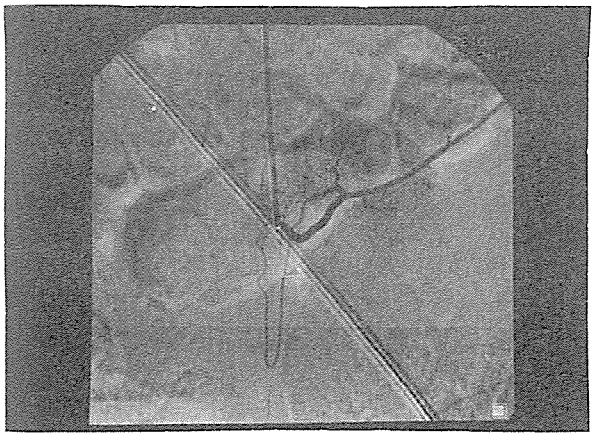

Fig. 4. Control angiogram of right D11 intercostal artery confirms the persistance of occlusion and patency of the anterior spinal axis. It is now remodelled and retained a normal size.

DISCUSSION

Juvenile AVM's are rare lesions and formerly known as Type III AVM's. They affect children, adolescents and young adults with equal incidence in two sexes especially in cervical and upper thoracic region (8, 9).

Majority of the patients with Juvenile AVM's have pain and acute neurologic dysfunction caused by subarachnoid or intramedullary haemorrhage due to high–flow lesion. Rapid blood flow may frequently cause arterial and venous aneurysms and subarachnoid or intramedullary haemorrhage may attribute to these lesions. Other patients have a gradual onset and progression of symptoms resulting from watershed infarction due to arterial ischemia by a vascular steal phenomenon or a result of venous congestion. Arterial aneurysms and/or venous varices may also cause mechanical cord compression by spontaneous thrombosis and mass effect. The combination of these pathologies are arterial steal, cord ischaemia, and progressive myelopathy in patients with juvenile AVM's who do not have haemorrhage.

Cerebral AVM (14), ankylosing spondylitis (3, 7), Klippel–Trenaunay–Weber syndrome (7) are associated anomalies with spinal AVM's.

Migraine (10), meningitis (6), abdominal pain (2), intracranial subarachnoid haemorrhage (5) are unexpected presentations of spinal AVM's.

They occupy the entire spinal canal especially ventral half of the spinal cord at longitudinal direction. Juvenile AVM's are always high–flow, complex lesions and involve intramedullary, extramedullary and even extraspinal areas. Feeding arteries are generally multiple and originate from dilated posterolateral and especially anterior spinal arteries. MRI and angiography are the most useful imaging modalities for the diagnosis. Subarachnoid haemorrhage may be seen during angiography and/or embolization. Similar to our case, Sato et al. have published a case with spinal AVM that subarachnoid haemorrhage occurred following the injection of contrast material after insertion of catheter into the feeding artery (12).

During surgery; feeding arteries are generally not recognized from enlarged spinal arteries that also supply the spinal cord. Normal spinal cord neural tissue is seen within the AVM and unfortunately most of these lesion can not be safely excised. Therefore

these lesions are generally considered unresectable. Rare case reports of successful excision of juvenile AVM's following embolization that are located dorsal half of the spinal cord have been presented (13). In our case, a bled tumor was thought primarily due to low thoracic location of lesion. A juvenile type AVM was found at operation, but feeding arteries could not be found in spite of the many–sided test occlusion by bipolar forceps. Any discoloration or haematoma also could not be performed because of the probable presence of anterior and multiple feeding arteries.

Embolization with embolic material such as isobutyl–cyanoacrylate or polyvinyl alcohol (Ivalon) is the recommended treatment in patients with juvenile AVM's (2). Frequently, feeding arteries of the AVM can not be obliterated permanently while preserving the blood supply of the normal cord in the first embolization due to origin of these arteries from the medullary arteries. Although the embolization does not seem to be the radical treatment, it should be repeated to avoid progressive myelopathy and to reduce the risk of subsequent haemorrhage by decreasing blood flow of these high–flow lesions. It is also possible that aneurysms in the nidus present high risk for rehaemorrhage as observed in brain AVM's. After a year's follow–up, it can be concluded that occlusion of aneurysms can prevent rebleeding as observed in the present case. Thus, embolization yields improvement as well as stabilization of neurologic functions and reduces the rebleeding rate.

CONCLUSION

Juvenile arteriovenous malformations are always high–flow lesions and their blood supply is from multiple medullary arteries via the anterior and/or posterior spinal arteries. Feeding arteries are generally not recognized from enlarged spinal medullary arteries that also supply the spinal cord. These lesions are generally considered unresectable. Embolization is the recommended treatment in patients with progressive myelopathy or haemorrhage.

REFERENCES

1. Barrow DL, Colohan ART, Dawson R: Intradural perimedullary arteriovenous fistulas (Type IV spinal cord arteriovenous malformations). J Neurosurg 81: 221–229, 1994.
2. Bjork A, Eeg–Olofsson O, Svendsen P, Mostrom U, Pellettieri L: Endovascular treatment of a spinal arteriovenous malformation in a 21–month–old boy. Acta Peditr 83: 1326–1331, 1994.
3. Chen JY, Ho HH, Wu YJ, Luo SF: Coexistence of spinal arteriovenous malformation and ankylosing spondylitis, Are they related? Clin Rheumatol 13: 533–536, 1994.
4. Heros RC, Debrun GM, Ojemann RG, et al: Direct spinal arteriovenous fistula: A new type of spinal AVM. Case report. J Neurosurg 64: 134–139, 1986.
5. Ikeda H, Fujimoto Y, Koyama T, Fujimoto Y: A rare case of high cervical spinal cord dural arteriovenous fistula presenting with intracranial subarachnoid hemorrhage. No Shinkei Geka 22: 1045–1048, (abstract) 1994.
6. Lowe DW, Gospe SM Jr, Hecht ST: Spinal arteriovenous malformation presenting as meningitis. Dev Med Child Neuro 38: 549–553, 1996.
7. Moritoyo T, Hashimoto K, Furuzono H, Nagamatsu K: A case of Klippel–Trenaunay–Weber syndrome, special reference to myelopathy. Rinsho Shinkeigaku 33: 746–753 (abstract) 1993.
8. Muraszko KM, Oldfield EH: Vascular malformations of the spinal cord and dura. In: Stein BM, McCormick PC (Eds). Neurosurgery Clinics of North America. Vol 1/No. 3, pp. 631–652, Philadelphia, WB Saunders: 1990.
9. Oldfield EH: Spinal vascular malformations. In: Rengachary SS, Wilkins RH (Eds). Neurosurgical operative atlas, Vol. 4, pp. 1–18, Illinois, AANS Publication Committee: 1995.
10. Ravishankar K: A ruptured spinal AVM in a patient with migraine (letter). Cephalgia 16: 280. 1996.
11. Rosenblum B, Oldfield EH, Doppman JL, et al: Spinal arteriovenous malformations: A comparison of dural artetiovenous fistulas and intradural AVM's in 81 patients. J Neurosurg 67: 795–802, 1987.
12. Sato M, Yamada R, Nishida N, et al: Pitfall of superselective catheterization by 3F catheter for spinal arteriovenous malformation. Radiat. Med. 13: 259–261, (Abstract) 1995.
13. Touho H, Karasawa J, Shishido H, Yamada K, Shibamoto K: Successful excision of a juvenile–type spinal arterivenous malformation following intraoperative embolization. Case report. J Neurosurg 75: 647–651, 1991.
14. Tsurushima H, Meguro K, Matsumra A, Narushima K, Nakada Y, Nose T: Multiple arteriovenous malformations of spinal cord and brain in a child. Pediatr Neurosurg 23: 166–170, 1995.
15. Waters RL, Adkins RH, Yakura JS: Definition of complete spinal cord injury. Paraplegia 9: 573–581, 1991.

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