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Case Report

Intradural Arteriovenous Malformation Located at Conus Medullaris: A Case Report and Literature Review

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Abstract: Arteriovenous Malformationis a rare primary lesion of the spine. The lesion could be located extra or intra dural. The conus medullaris malformation is a special lesions which leads to progressive myelopathy or radiculopathy. The lesion is characterized by multiple feeding arteries, multiple niduses, and complex venous drainage. The neurological deficits may be improved dramatically over time when these lesions are successfully treated, however, the numerous feeding arteries and multiple niduses create a potential for recurrence.We report a case of a 36-year-old man complaining of serious lower limbs muscle weakness. The MRI scan showed multiple arteriovenous systems go around the conus medullaris. The spinal cord is enlarged with spinal cord edema. Embolization was proformed to stop the blood flow of main feeding artery. Post-operation the lower limb muscle weekness has been significantly improved. The patient can walk without additional support. Until the 3 months follow-up, the clinical result was satisfied.

Keywords: Arteriovenous malformation; spine; intramedullary.

INTRODUCTION

Spinal arteriovenous malformations (AVMs) are reported as the second most common spinal vascular malformation, constituting up to 15% of all spinal vascular malformations [1]. The anterior spinal artery and posterior spinal artery typically provide the arterial supply for the intramedullary or perimedullary nidus, which flows into a medullary vein [2, 3]. The conus medullaris arteriovenous malformations is characterized by multiple feeding arteries, multiple niduses, and complex venous drainage. They have multiple direct arteriovenous shunts that derive from the anterior and posterior spinal arteries and have glomus-type niduses that are usually extra-medullary and pial based, but they may also have an intra-medullary component [4, 5]. They can extend along the entire terminal filum. Symptomatically, they can manifest with venous hypertension, compression, or hemorrhage, producing radiculopathy and myelopathy [6].

CASE REPORT

History

A 36-year-old man with history of 2 months of gradual onset of progressing lower limb weakness presented to our department. The muscle weakness can not be relieved by any spinal movement or posture. Bed rest could not improve the symptom. He has consulted medical attention at local hospital, and received MRI exam. The result reviewed a vascular malformation located at the conus medullaris. He was refer to our department when he could hardly walk. He began to complain urinary retention and bowl movement problems. Another MRI scan with enchantment was performed. The detail structure of the vascular malformation had been reviewed. he denied any other systemic diseases. He was conscious and did not have any difficulty of communication.

Examinations

Physical examination revealed decreased motor and sensory function below groin of both sides, the impairment of muscle tension was the major problem of walking. The manual muscle strength test demonstrated 3/5 strength on the left and 3/5 on the right. Increase of deep tendon reflex was noted. Babinski sign was positive bilaterally. Muscle tonus of lower extremities was significantly increased.The dysuria function was abnormal. X-ray images showed no pathological destruction and loss of spinal column support. MRI scans revealed no spinal stenosis, lumbar disc herniation or never root compression. However, the conus medullaris was enlarged with edema. The lesion consisted of multiple fistulas feeding the malformation located around surface of the conus. The mass was hypointense on T1-weighted images and hyperintense on T2-weighted images with marked high signal on STIR images. A major feeding artery was discovered in

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MRI angiography. Laboratory examinations consisted of complete blood count, electrolytes, liver, renal function tests, urinalysis. They were all normal (Fig 1 and 2).

Fig-1: Pre-operative STIR MRI pictures

Fig-2: Fast 3D contrast-enhanced MRA

Surgical Treatment

The main feeding artery were considered to be responsible for clinical symptoms. Embolization was performed to stop the blood flow. The embolization procedures were performed using local anesthesia and neurolept analgesia with frequent testing of motor and sensory functions. A double femoral artery puncture was performed. One vascular approach was used to introduce the embolic agents and the other was used to control the other arterial feeders. Once the embolic agents was introduced into the target area, complete occlusion of the shunt was confirmed by angiograms obtained via the control catheter. Patients with feeding arteries close to the artery of Adamkiewicz or the anterior spinal artery and a small lumen at the origin of the target vessel may be at high risk for spinal cord infarction due to migration of the embolic material [7]. Open surgical was prepared if there was worsening of clinical symptoms.

Postoperative Course

One weeks later, his muscle strength was improved. The manual muscle test demonstrated 4/5 strength on the left limbs and 4/5 on the right limbs. After rehabilitation, the patient could walk independently for 10 meters. He was discharged home 2 weeks later. After follow-up for 3 months, neurology examination revealed decreased muscle tonus of lower extremities, there was no recurrence of muscle weakness.

The design and performance of this study conformed to ethical standards of Helsinki Declaration and our national legislation. It was approved by Medical Ethical Committee of our institution. The patient was enquired whether or not willing to take part in a scientific report and informed consent forms were signed by himself.

DISCUSSION

The literature on spinal vascular malformations contains a great deal of confusing terminology. Some of the nomenclature is inconsistent with the lesions described. Based on a thorough review of the relevant literature, Lesions could be defined into three primary or broad categories: neoplasms, aneurysms, and arteriovenous lesions. Neoplastic vascular lesions include hemangioblastomas and cavernous malformations, both of which occur sporadically and familially. The second category consists of spinal aneurysms, which are rare. The third category, spinal cord arteriovenous lesions, could be divided into arteriovenous fistulas and arteriovenous malformations (AVMs). Intradural AVMs are further divided into different categories according to their location. The conus medullaris is a new category of AVM [3-5].

The conus medullaris malformationis a special lesions which leads to progressive myelopathy or radiculopathy. The lesion is characterized by multiple feeding arteries, multiple niduses, and complex venous drainage [6]. They have multiple direct arteriovenous shunts that derive from the anterior and posterior spinal arteries and have glomus-type niduses that are usually extra-medullary and pial based, but they may also have an intra-medullary component. They can extend along the entire terminal filum. Symptomatically, they can manifest with venous hypertension, compression, or hemorrhage, producing radiculopathy and myelopathy [7-9].

Currently, the most commonly used MRA technique for detecting and evaluating spinal AVM is fast 3D contrast-enhanced MRA using elliptic centric k-space encoding with an acquisition time less than 1 minute/phase and spatial resolution adequate for demonstrating pathological intra-dural vessels [10, 11]. Our spinal MRA technique has 2 different technical aspects, which are 1) the use of hand syringe contrast injection instead of an automatic contrast injector machine and 2) the dose of the gadolinium-based contrast agent lowered to 0.15 mmol/kg. We have showed that limiting the use of contrast media, not using an automatic injector machine, and using our fast 3D contrast-enhanced MRA technique with manual syringe contrast injection can accurately detect the presence of spinal AVM, locate the level, and discriminate the AVM subtype in most cases, but is limited when detecting small arterial feeders. As you can see, we have received nice pictures of the detail structure of the conus medullaris AVMs.

Conus AVMs can be treated with both surgery and embolization [12, 13]. Our experience with this type of lesion indicates that definitive treatment is best achieved using aggressive embolization followed by subsequent resection. If there is neurological recovery after embolization, in-patient observation should be done for a week. If there was no more deteration, the patient could be discharged. Arteriovenous malformations of the conus can be challenging to treat especially for asymptomatic or mildly symptomatic lesions. Preoperative mapping of the anatomy usually requires a catheter angiogram, and intraoperative study of the lesion can be done non-invasively using angiography. Embolization can carry significant risk compared to cranial vascular malformations because of the terminal nature and the caliber of vessels of the spinal cord [14]. Neuromonitoring can be useful especially with lesions encroaching on the cord or the conus itself.

CONCLUSIONS

Although the presentation of radiculopathy and myelopathy are commonly seen in clinical practice, spinal arteriovenous malformations (AVMs) located at

the conus medullaris is special in its numerous feeding arteries and multiple niduses. Fast 3D contrast-enhanced MRA is useful in discovering the detail structure of the lesion. Since the recurrence rate is high, the treatment should be aggressive embolization followed by subsequent resection. However, neurological deterioration was not observed in the reported patient after embolization, and close follow-up is conducting.

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