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Multiple Intramedullary Cavernous Angioma of the Cervicothoracic Spine: Case Report and Review of the Literature

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Abstract Intramedullary cavernous angioma (ImCA), which was originally thought to be an uncommon vascular lesion, has shown an increased incidence since the advent of magnetic resonance imaging. We present a case of multiple ImCAs in the cervicothoracic region. The patient presented with slow, progressive motor weakness in the upper and lower limbs. Surgical intervention was performed because of misdiagnosis as a spinal cord tumor. The patient underwent decompression surgery because no neoplasm was noted in the course of the operation. The angioma regressed on follow-up magnetic resonance images 3 months later.

Keywords: *multiple intramedullary cavernous angiomas, spinal cord, cervicothoracic segment, decompression surgery, magnetic resonance imaging*

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1. Introduction

Intramedullary cavernous angiomas (ImCAs) are usually solitary, but may be multiple, with similar lesions affecting other organs or the central nervous system. These lesions are angiographically occult malformations because they have no large arterial supply or major venous drainage. ImCAs are characterized by sinusoidal vascular channels, which are lined by thin endothelial cells without intervening normal nervous tissue. Bleeding of ImCAs around blood vessels causes hemosiderin deposition in the surrounding neural tissue [1]. The most common clinical presentation of ImCAs is a slowly progressive myelopathy. The typical characteristics of ImCA have been well described in the literature [1,2,3].

We report a 39-year-old female with multiple ImCAs for which we performed decompression surgery. The patient underwent clinical observation after surgery and the lesions regressed on follow-up MRI 3 months later.

2. Case Presentation

A 39-year-old woman presented with neck pain and weakness in the upper and lower extremities, and these were worse on the right side. This complaint had begun 5 days before admission and progressively increased. A clinical examination showed sensory deficits on the perineal region and painful dysesthesia below the bilateral supraclavicular fossa. A neurological examination showed hyperreflexia of the bilateral triceps muscles, biceps muscles, and right Achilles tendon. Bilateral Hoffmann's signs and Babinski's signs were observed. This study was approved by the Institutional Review Board of our hospital.

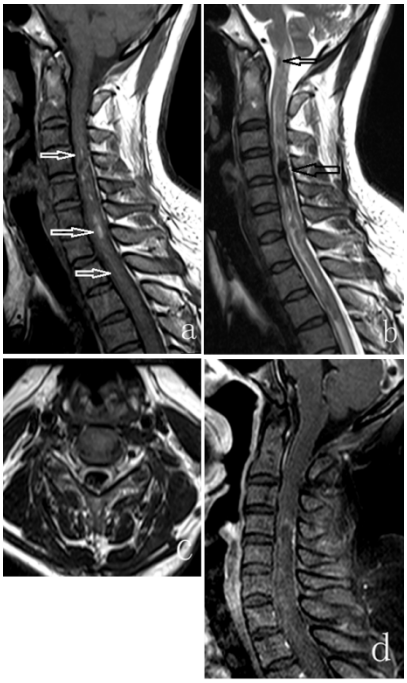

Figure 1. Multiple intramedullary cavernous angiomas in a 39-year-old woman. a. Multiple hyperintense signals represent intralesional hemorrhage at the level from C4 to C7 on a sagittal T1-weighted image (arrow). b. The dark area represents hemosiderin deposition (arrow) on a sagittal T2-weighted image and is associated with extensive edema. c. One lesion occupies almost the entire spinal cord on an axial T2-weighted image. d. Postcontrast T1-weighted sagittal MR image showed multiple patchy hyperintense signal in the spinal cord

A full blood count and biochemical test results were within the normal limits. We used an American GE Signa HDX 1.5 T superconducting magnetic resonance scanner in our case. MRI showed multiple, discrete, intramedullary lesions extending at the level from C1 to T2. Multiple lesions showed high-intensity signals relative to the spinal cord, representing intralesional hemorrhage on T1-weighted images. The hemorrhage appeared as an isointense signal on T2-weighted images. However, there was extensive intramedullary edema, which showed a hyperintense signal on T2-weighted images. The largest lesion measured 23 mm in the maximum superior-inferior diameter and occupied almost the entire spinal cord. The largest lesion was a heterogenous hypo- and hyperintense signal on T1- and T2-weighted images, representing hemorrhage mixed with hemosiderin (Figure 1a–c). Enhanced MRI was obtained using a T1-weighted spin-echo pulse sequence after administering gadolinium. The sequence was as follows: repetition time/echo time, 500/5.5 ms; slice thickness, 3.0 mm; interslice gap, 0.5 mm; field of view, 22 cm × 22 cm; and matrix size, 320 × 192. There were multiple patchy areas of enhancement in the spinal cord (Figure 1d).

Our patient underwent C5 laminectomy. After a midline vertical skin incision in the posterior part of the neck, the paravertebral muscle was dissected bilaterally and the spinal processes of cervical vertebrae were exposed. Laminectomy at the level of C5 was performed and the dura was opened under magnification by microscope. Myelotomy was performed via the posterior median fissure and there was no neoplasm noted in the course of surgical intervention. The patient did not undergo microsurgical excision because the lesions were deep-seated and decompression surgery was performed. The patient underwent clinical observation after surgery. At a 3-month follow-up, a spinal MRI scan showed that swelling of the spinal cord was improved, and some lesions had considerably regressed (Figure 2e–g). The clinical presentation of the patient was greatly improved.

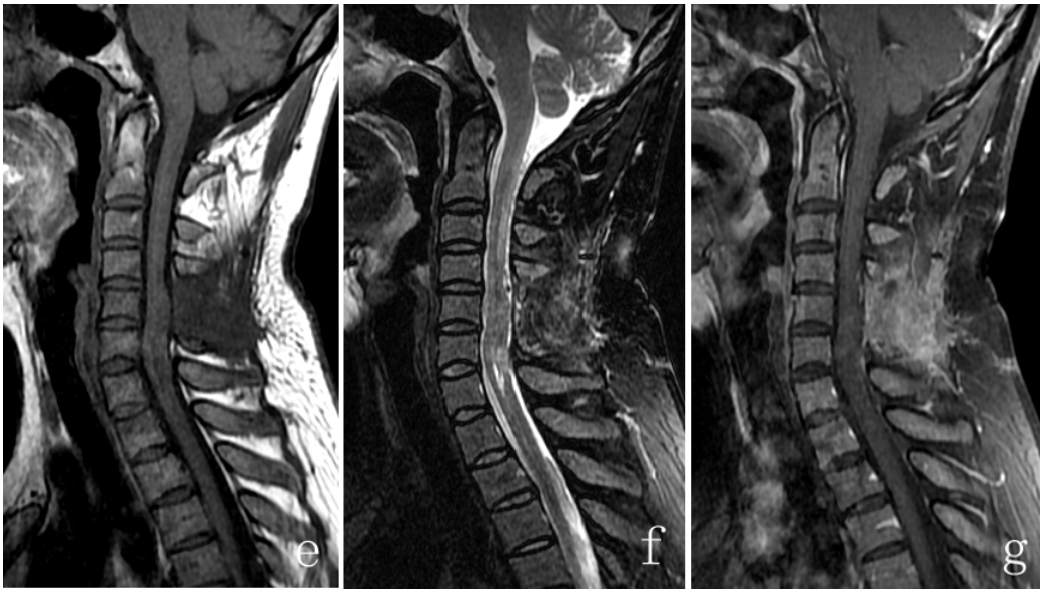

Figure 2. Postoperative MRI follow-up 3 months later. Postoperative sagittal T1-weighted image (e), T2-weighted image (f), and contrast T1-weighted image (g). Hemorrhage and hemosiderin deposition are completely resolved, with partial resolution of the edema. There is no obvious enhancement on T1-weighted, gadolinium-enhanced, sagittal MRI

3. Discussion

The entity of spinal ImCA was first reported in 1903 [2]. According to Lee et al. [3], the incidence of cavernous angioma involving the central nervous system is from 0.49% to 0.53%. ImCAs account for approximately 12% of spinal cord vascular lesions [3]. The thoracic spinal cord is affected in more than half of cases [4].

ImCAs may be found in all organs of the human body [5]. A total of 75% of individuals with multiple lesions in the spinal cord are familial and are associated with an autosomal dominant genetic defect. Multiple lesions are typically seen in so-called inherited cavernous angioma. These mutations include CCM1 on chromosome arm 7q, CCM2 on 7p, and CCM3 on 3q, which may account for approximately 40–50%, 10–20%, and 40% of inherited cases, respectively [6]. Approximately 10% to 25% multiple ImCAs are sporadic cases, and the remainder of multiple ImCAs in individuals may be the result of secondary effects of radiation therapy [7].

Generally, the clinical presentation of ImCAs is variable, often including sensory and motor symptoms, resulting in pain and paresis. Several factors may contribute to spinal cord injury following hematomyelia. Ischemic damage may occur because of a decrease in blood flow. Vasospasm could occur as a result of spinal cord exposure to red blood cells and inflammatory mediators. Vasogenic edema can result from damage of the blood–spinal cord barrier [8]. Deposition of hemosiderin may alter the surrounding microcirculation, which affects progression of symptoms in patients who do not show evidence of enlargement or re-bleeding of angiomas on MRI [9]. Occasionally, the symptoms may recover because of repeated microhemorrhage from an angioma followed by gliosis or iterative thrombosis within the lesions [8].

MRI is the gold standard for diagnosis of ImCAs in any location. ImCAs typically appear as small, ovoid well-

defined, “popcorn-like” lesions with mixed signal intensity on MR scans because of blood products in various stages of evolution. A rim of low-signal intensity around a hyperintense core lesion is caused by hemosiderin deposition around the cavernoma on T2-weighted images. When ImCAs are followed up conservatively without surgical intervention, intramedullary edema and hemorrhage can disappear, as well as residual blood products around the ImCAs [9]. However, spinal cord re-bleeding causes deterioration, which leads to a swollen spinal cord with cord edema.

The current treatment of symptomatic or minimal symptomatic ImCAs consists of total excision with a microsurgical technique [10]. The hemorrhage rate of unruptured spinal cord cavernomas is estimated to be 1.4% per lesion and year. Mechanical distortion and tissue plasminogen factor might play large roles in bleeding of ImCAs [11,12]. Grasso et al. [9] reported that a subtotal excision should be advocated in symptomatic patients. However, a high incidence of re-bleeding in partially excised cavernomas ranges from 9% [11] to 18% [12] of cases. Al-Khuwaitir et al. [13]described that surgical resection of symptomatic lesions should be advocated to prevent intramedullary hemorrhage and halt progressive neurological decline. Once the lesion has been removed, the cavity must be carefully inspected to identify any residual malformation that may lead to future re-bleeding and recurrence of symptoms. Badhiwala et al. [14] reviewed neurological outcomes for 632 patients with ImCAs who underwent operative or conservative (observation) treatment. They showed that surgery in these patients was better than conservative management. Patients will benefit from resection of the lesion within 3 months of onset of symptoms, and hemilaminectomy is better than total laminectomy or laminoplasty. Furthermore, the rate of neurological improvement for patients who had surgical resection performed was no higher than that for patients with superficial cavernous malformations or for those with deep-seated or ventrally located lesions.

ImCAs are relatively uncommon, but are a surgically curable condition of myelopathy. Early diagnosis and proper management can prevent hemorrhage and dangerous enlargement of lesions. Most lesions regress and there is partial resolution of hemosiderin. Clinical representation improved may resulting from decompression surgery in our case of multiple ImCAs.

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