CASE REPORT

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Isolated Hemangioblastoma of the Filum Terminale

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The filum terminale is an exceptional location for isolated hemangioblastoma, and most commonly hemangioblastomas are present in patients with von Hippel-Lindau(VHL) syndrome. We describe here a case of hemangioblastoma of filum terminale not associated with VHL, presenting with the history of progressive back pain, particularly severe in recumbent posture, and recurrent bilateral sciatica. MRI and spinal angiography revealed a well-vasculized mass lesion in filum terminale. The tumor was resected surgically. Histological examination confirmed the hemangioblastoma diagnosis. We recommended that, although rare, hemangiblastoma of the filum terminale be included in the differential diagnosis of a patient with low back pain.

KEY WORDS : Hemangioblastoma · Filum terminale · von hippel-Lindau syndrome.

Introduction

Hemangioblastomas of the spinal cord is not a common lesion, and those located at the filum terminale are very rare. Spinal cord hemangioblastoma accounts for 1.6~5.8% of all primary spinal tumors[2,16] and for 11~17% of central nervous system hemangioblastoma[1,2,9]. Most commonly these tumors are present in patients with von Hippel-Lindau(VHL) syndrome. The association between the lesion and VHL disease is detected in approximately 30~64% of the cases[2,5,10], but mutations of the VHL gene are demonstrated in about 80% of patients with an apparently sporadic tumour[10]. We describe here a case of a 42-years-old man with a pure hemangioblastoma, not associated with VHL.

Case Report

Our patient was a 42-year-old healthy man with a 7-year history of low back pain. He noted recent worsening of the pain, particularly severe when in recumbent position, and recurrent bilateral sciatica. Attempts of bed rest and administration of non-steroidal anti-inflammatory medications did not result in relief of the pain. The patient experienced no change in bladder or bowel function and had no history of low back surgery or lumbar punctures. Motor and sensory examinations of both lower extremities were unremarkable. Also no other abnormal neurological signs were present.

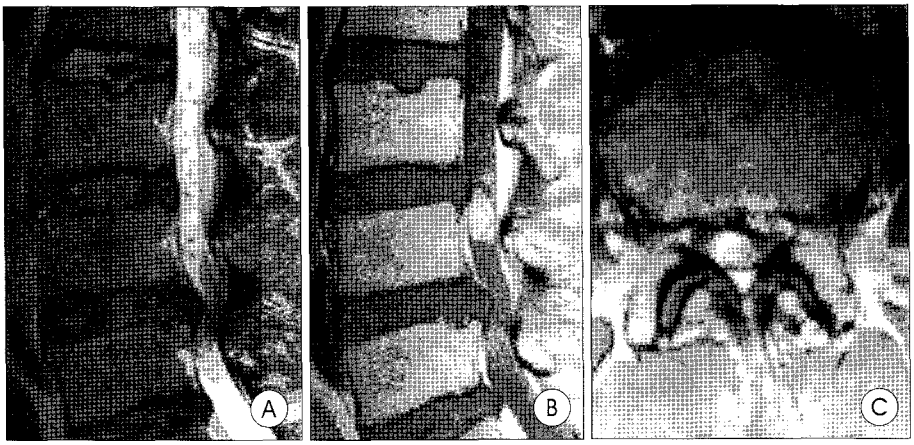

Fig. 1. On magnetic resonance images, sagittal T2 weighted sequence shows multiple vascular signal voids seemed to be abnormally dilated serpiginous vessels (A). On enhanced sagittal T1 weighted sequence the tumor appears to be homogeneous intradural enhancing mass with Gadolinium-DPTA-dimeglumine contrast at the L4 level (B). Axial T1 weighted sequence with contrast shows enhancing mass occupying most of the spinal canal (C).

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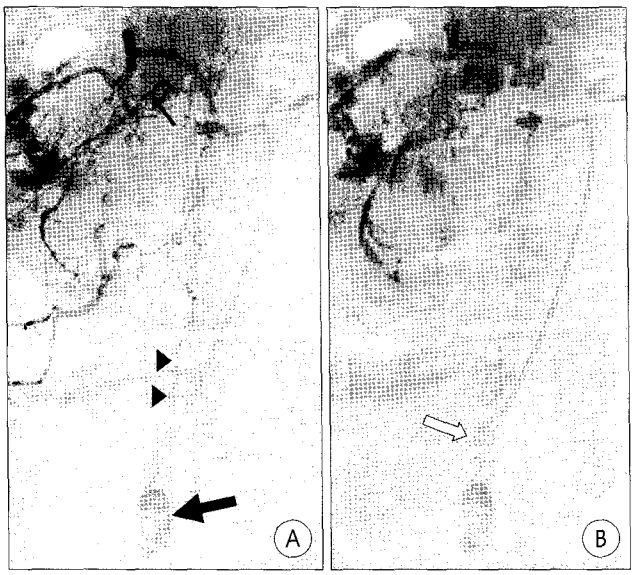

Fig. 2. On spinal angiography, the mass has homogeneous dense blush (large arrow) from posterior spinal artery (arrow heads) contributed from dilated right radicular artery (small arrow) of L1 segmental artery on the arterial phase (A) and early venous drainage (white arrow) on venous phase (B) which seems to be multiple signal voids on magnetic resonance image.

Fig. 3. Intraoperative photography shows the beefy red hemangioblastoma with large artery and vein (white arrow) arising from the upper portion of the tumor and an enlarged filum (black arrow) extending from the inferior pole of the tumor.

Only degenerative disc abnormalities were previously noted in lumbar CT, which was performed elsewhere without a contrast medium. Sagittal T2 weighted sequences showed multiple vascular signal voids seemed to be the abnormally dilated serpiginous vessels displayed above the rostral portion of the tumor as well as a disc protrusion at the L4-5 level (Fig. 1A). On enhanced sagittal T1 weighted sequences the tumor appeared as a homogeneous sharply defined intradural enhancing mass (16mm) with Gadolinium-DPTA-dimeglumine (Gd-DTPA) contrast in the L4 level (Fig. 1B). Axial T1 weighted sequences with contrast showed an enhancing mass occupying most of the spinal canal (Fig. 1C). On angiography, posterior spinal

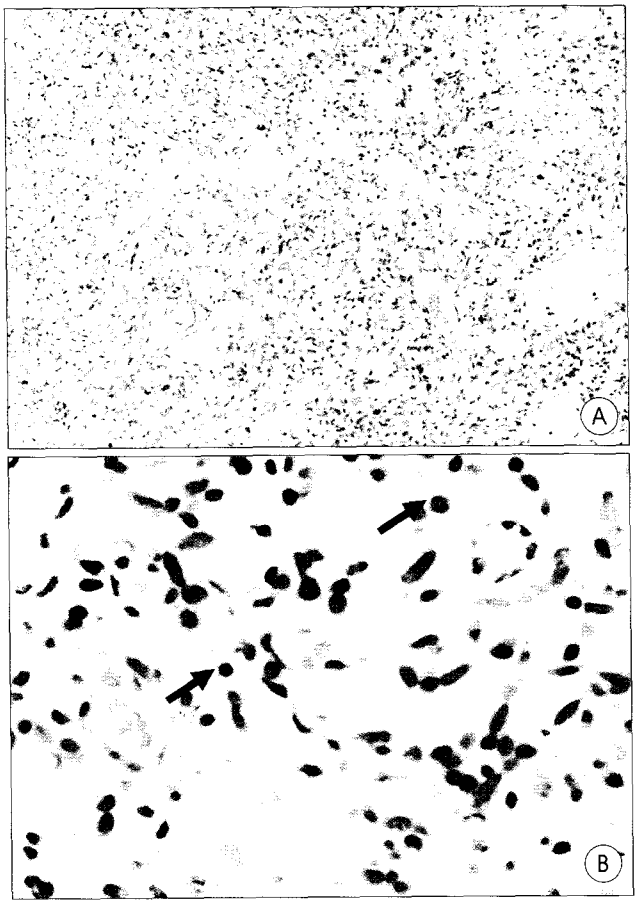

Fig. 4. Histopathologic findings showing a vascular neoplasm and complex capillary network (A). There are lipid-containing vacuolated stromal cells (arrows) in the interstices of a capillary network (B). (H&E : original magnification, A : ×200 ; B : ×400).

artery contributed from marked dilated right side radiculopial artery derived from left L1 segmental artery fed to the round shaped tumor mass at the level of L4 upper body. The mass had a homogenous dense tumor blush and early venous drainage which seemed to be multiple signal voids on MRI (Fig. 2). Investigations for VHL disease included serological test, computed tomography of the head and abdomen, and a neuro-opthamological examination were all normal.

An L3-L5 laminectomy was performed, confirming the presence of a reddish, solid and capsulated mass arising from the filum terminale and extending 1.7cm in length with overlying adherent nerve roots of the cauda. The tumor was richly vascularized and fed by posterior spinal artery. At the rostral pole draining dilated vein was showed. At the caudal pole an enlarged filum extended from the tumor, together with a feeding artery (Fig. 3). The filum was sectioned and the mass was completely removed.

The surgical specimen was beefy red soft tissue in appearance. A vessel pair was contiguous with the mass. Microscopically, the mass contained abundant fat-laden stromal cell, endothelial cells, pericytes, and a large artery, which revealed typical vascular

structure of hemangioblastoma (Fig. 4). Postoperative course was normal and pain greatly improved. Nine months after surgery, pain is occasional and the patient is fully employed. Repeated investigation for VHL disease remain negative and MRI demonstrate no evidence of recurrence.

Discussion

Farneti et al.[3] reported that spinal cord hemangioblastoma accounts for 1.6~5.8% of all primary spinal tumors, and the tumors are more frequently intramedullary (60~75%) and located in the thoracic and cervical levels in over 90% of the cases[2,5]. Tibbs et al.[13] presented that 21 to 28% of spinal cord hemangioblastoma located intradural-extramedullary and 9 to 13% of these found within the cauda equina. The association with von Hippel-Lindau is found in only 30% of all spinal hemangioblastomas[2,5]. Isolated hemangioblastomas of the filum terminale not associated with VHL are very rare.

We report the case of a 42-year-old man with a hemangioblastoma of the filum terminale. A literature review found only six additional cases. In 1943, Wyburn-Mason[14] presented the report of a 25-year-old woman with radicular pain in the L2-S2 distribution. Operative intervention yielded a hemangioblastoma of the filum terminale. Sloof et al. reported a patient with a hemangioblastoma of the filum terminale in his monograph on tumors in 1964[3,13]. In 1985, Wolbers et al.[15] published a case presenting CT images of a 36-year-old man who experienced lumbo-radicular pain. Silvermann et al.[11] examined a case in relation to ultrastructural and immunochemical features. Tibbs et al.[13] reported the case of a hemangioblastoma of the filum terminale and summarized previous reported three cases by review of literatures. Farneti et al.[3] emphasized that MR finding is pathognomonic for diagnosis of hemangioblastoma of filum terminale that was not noted in non-enhanced CT.

In these seven cases including our case, the range of age were from 25[14] to 66 years old[11]. Progressive back pain and sciatica were the most common symptoms shown in theses patients[3,14,15]. The symptoms in tumors of the cauda can be erroneously referred to spondylosis or disc prolapse[6]. Actually some patients had degenerative disc at the time of diagnosis[3,13]. Another common symptom was recumbent pain. This is of particular importance, unlike in disc prolapse[6]. This condition is probably due to an increased tension in the caudal nerve roots caused by a decreased lumbar lordosis occurring in recumbency. An alternative hypothesis could be raised cerebrospinal fluid pressure during sleep.

With regard to radiological diagnosis, in a case of hemangioblastoma of the filum, Wolbers et al.[15] noticed that CT performed without a contrast solution did not contribute to a specific diagnosis differently with a contrast CT. The interest of Gd-DTPA-enhanced MRI in spinal hemangioblastoma is largely emphasised in literature[12]. In 1989 Murota et al.[8], presenting 18 cases, stated that MRI provides an accurate demonstration of the mass and the feeding vessels. More recently Grunberg et al.[4], reviewing 20 patients, confirmed that MRI is the selected diagnostic modality and that angiography should not be used. Traditionally, however, this has not been the case, and spinal angiography remains the gold standard for preoperative diagnosis. Embolization is not necessary, but the ill-prepared surgeon will find him- or herself in trouble managing a heavily bleeding hemangioblastoma[13]. So Lee et al. recommended preoperative spinal angiography and embolization because it might prevent intraoperative bleeding and improve surgical outcome[7].

In the literature five cases of solitary hemangioblastoma of the film terminale, except the case of Silverman, were all operated on. It is necessary to remove theses tumors completely, because incomplete excision usually results in recurrence of symptoms, even if the tumor is treated with radiotherapy[2,16].

Conclusion

We experienced a rare case of hemangioblastoma in the filum terminale. The case indicates that hemangioblastoma, although uncommon, must be taken into consideration in cauda equina tumor diagnosis, and also emphasized the specificity of magnetic resonance features, and spinal angiography can aid in diagnosis.

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