CASE REPORT

Spinal Cord Hemangioblastoma with Extensive Syringomyelia

Te-Chang Wu[1], Wan-Yuo Guo[1], Jiing-Feng Lirng[1,2]*, Tai-Tong Wong[3], Feng-Chi Chang[1], Chao-Bao Luo[1], Michael Mu-Huo Teng[1,2], Cheng-Yen Chang[1]

[1]Department of Radiology, Taipei Veterans General Hospital, [2]National Yang-Ming University School of Medicine, and [3]The Neurologic Institute, Taipei Veterans General Hospital, Taipei, Taiwan, R.O.C.

We present the case of a 20-year-old male with intermittent right upper extremity numbness for 3 months. His pain perception and temperature sensation were severely disturbed. An incidental magnetic resonance imaging (MRI) finding of one small intramedullary enhancing nodule at spinal cord level T10–11 with long-segment syrinx formation suggested the diagnosis of spinal hemangioblastoma with syringomyelia. Surgical removal of the tumor and decompression of the spinal cord with opening of the syrinx were performed smoothly, and the pathology confirmed the diagnosis of spinal hemangioblastoma. Reviewing the literature, MRI is the examination of choice for spinal hemangioblastomas, and is helpful in preoperative planning and the differential diagnosis of spinal cord neoplasms and vascular lesions. [*J Chin Med Assoc* 2005;68(1):40–44]

Key Words: hemangioblastoma, spinal cord neoplasm, syringomyelia, von Hippel-Lindau disease

Introduction

Hemangioblastoma is a common posterior fossa tumor in adults, but it is a relatively rare tumor of the spinal cord, accounting for 1–5% of all spinal cord tumors.[1–3] Here, we present a case of spinal hemangioblastoma with extensive syringomyelia and typical features on magnetic resonance imaging (MRI) and spinal angiograms. The literature is reviewed, and radiologic appearances of spinal hemangioblastomas and the pathogenesis of syrinx formation in spinal hemangioblastoma are discussed.

Case Report

A 20-year-old male had intermittent right upper extremity numbness for 3 months. His pain perception and temperature sensation were severely disturbed. At first, electromyography revealed suspected right C6–7 radiculopathy. Whole-spine MRI showed an intramedullary enhancing nodule, about 10 mm in size, in the right anterior portion of the spinal cord at level T10–11 (Figure 1), and a long segment of syrinx with multiple internal septa from the cervicomedullary junction to level T11 (Figure 2). High signal change of the spinal cord from T12 to the tip of the conus medullaris was also noted on T2-weighted images (T2WIs), indicating edematous change of the cord parenchyma (Figure 1B). Spinal angiograms showed a 15 mm hypervascular tumor, fed by a radiculopial artery arising from the right T12 intercostal artery (Figure 3). No family history of von Hippel-Lindau disease (vHLD) was noted, and brain MRI showed negative findings. Under the impression of an intramedullary tumor with extensive syringomyelia, laminotomy over T9–T12 was performed and showed an intramedullary vascular lesion at the right anterolateral aspect of T10–11 of the spinal cord, with a long segment of syrinx and cord swelling (Figure 4). Total removal of the tumor and decompression of the cord with syrinx opening were performed smoothly. The pathologic diagnosis was hemangioblastoma. The postoperative condition was uneventful, and the pa-

*Correspondence to: Dr. Jiing-Feng Lirng, Department of Radiology, Taipei Veterans General Hospital, 201, Section 2, Shih-Pai Road, Taipei 112, Taiwan, R.O.C.
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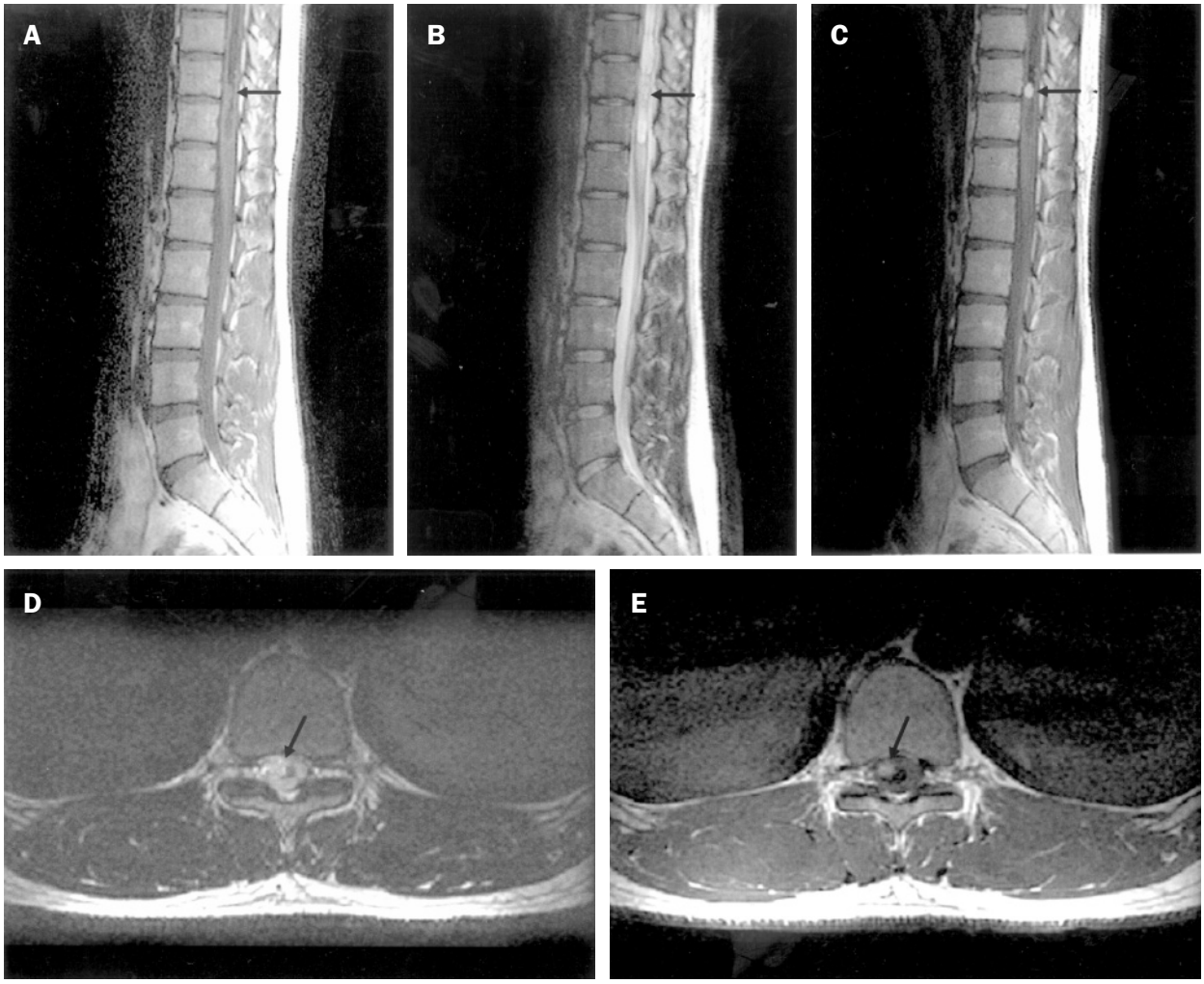

Figure 1. Fat saturation magnetic resonance imaging scans of the lower thoracic and lumbar spine showed an intramedullary enhancing nodule (arrow), about 10 mm in size, in the right anterior portion of the spinal cord at level T10–11: (A) sagittal T1-weighted image (T1WI); (B) sagittal T2-weighted image (T2WI); (C) post-gadolinium-enhanced sagittal T1WI; (D) axial T2WI; (E) axial T1WI. High signal change of the spinal cord from T12 to the tip of the conus medullaris was also noted on T2WIs, indicating edematous change of the cord parenchyma (B).

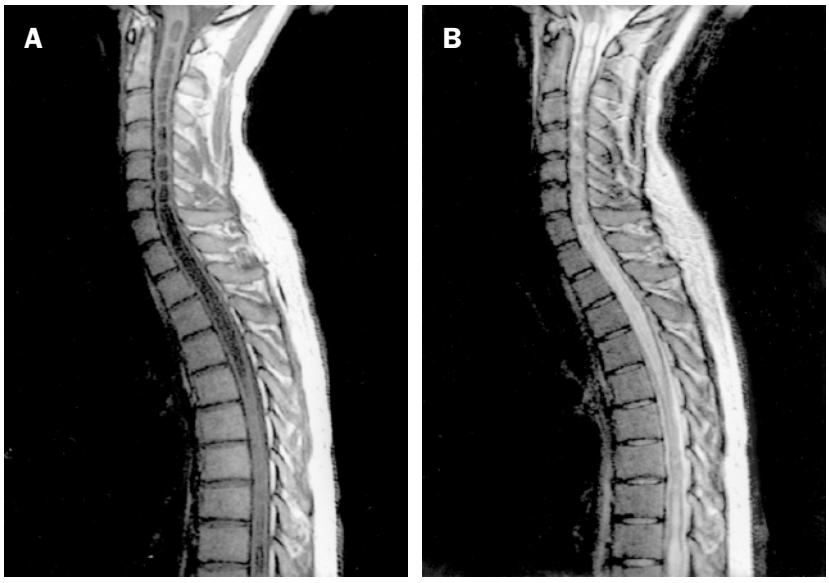

Figure 2. Magnetic resonance imaging scans of the C1–T9 spine showed a long segment of syrinx with multiple internal septa from the cervicomedullary junction to level T11: (A) sagittal T1-weighted image; (B) sagittal T2-weighted image.

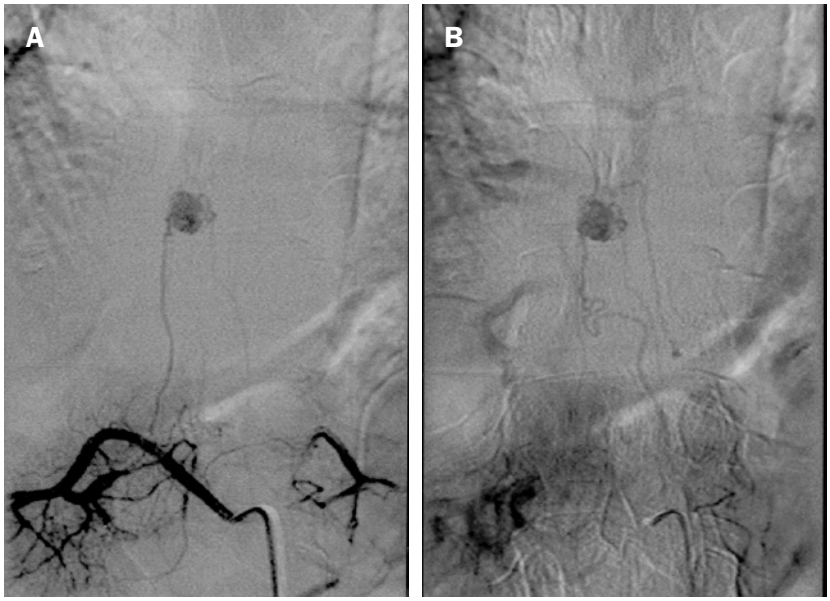

Figure 3. Spinal angiograms showed a 15 mm hypervascular tumor, fed by a radiculopial artery arising from the right T12 intercostal artery: (A) early arterial phase; (B) capillary phase.

tient received regular follow-up at our neurosurgery outpatient clinic.

Discussion

Hemangioblastoma, after ependymoma and astrocytoma, is the third most common intramedullary tumor of the spinal cord.[1,4] It may occur sporadically or be a component of vHLD. According to a review of histologically documented case reports by Browne et al, 32% of patients with spinal hemangioblastomas also had vHLD, and the remaining patients had sporadically occurring tumors.[5] About 80% of spinal hemangioblastomas are solitary tumors. Patients with vHLD, however, often have multiple hemangioblastomas and, indeed, such multiplicity of tumors indicates vHLD.[1–3] Otherwise, there are no differences in tumor imaging findings for sporadic and familial forms of spinal hemangioblastomas.[3] About 50–70% of spinal hemangioblastomas have been associated with syringomyelia.[1–3,5,6] This rate increases to 100% in some reports, when only intramedullary hemangioblastomas are considered.[7]

Figure 4. Laminotomy over T9–T12 was performed and showed an intramedullary vascular lesion at the right anterolateral aspect of T10–11 of the spinal cord with a long segment of syrinx and cord swelling.

Since its advent, MRI has been the examination of choice for spinal hemangioblastoma, and several characteristic MRI findings have been reviewed. Generally, small tumors (< 10 mm) are primarily isointense on T1-weighted images (T1WIs) and hyperintense on T2WIs. Larger tumors tend to be hypointense, or mixed hypointense and isointense, on T1WIs, and of heterogeneous intensity on T2WIs.[2,3] Small tumors frequently show well-demarcated intense enhancement, whereas larger tumors show heterogeneous enhancement.[2–4] Such an enhancement pattern represents a highly vascular tumor parenchyma comprising closely packed blood vessels interspersed with stromal cells.

The second characteristic MRI finding of spinal hemangioblastoma is the presence of "flow-void" phenomenon.[2–4] This is usually seen in larger tumors and reflects prominent feeding arteries or drainage veins, which are well demonstrated by spinal angiography. In a study of 12 patients with 32 spinal hemangioblastomas, flow-void appearance was found in seven patients (58%), and was invariably present when the tumor was larger than 25 mm, but not evident when the tumor was smaller than 10 mm.[3]

Hence, these investigators suggested that a diagnosis of spinal hemangioblastoma was unlikely when the tumor was ≥ 25 mm, and when vascular flow voids on MRI were absent. Sometimes, superficial enhancement of the spinal cord can be seen and confirmed as dilated perimedullary veins at surgery. Among small tumors, this is a rare MRI presentation, despite the possibility of abnormal distended vessels on spinal angiography.[3] In our presented case, there was vague superficial enhancement in the caudal portion of the tumor (not shown) that was not pointed out until comparison with spinal angiography.

The third characteristic MRI finding of spinal hemangioblastoma is superficial location of the intramedullary tumor, most often at the posterior aspect of the spinal cord.[3] This finding usually correlates with the surgical finding of subpial tumor location. The fourth characteristic MRI finding is the large size of the syrinx formation relative to the small size of the intramedullary portion of the tumor.[2–4] In our case, the long segment of syrinx from the cervicomedullary junction to level T11 was attributed to one small, 10 mm tumor nodule. Syrinx formation is not specific for spinal hemangioblastoma and can be associated with other spinal cord tumors, such as ependymoma and astrocytoma. However, the MRI presentation of one small, superficial, intramedullary nodule with intense enhancement and extensive syrinx formation is characteristic of spinal hemangioblastoma.[2–4]

The pathogenesis of syringomyelia has traditionally been unclear. However, numerous experimental studies have established that the subarachnoid space (SAS) and extracellular space (ECS) are 2 parts of a single fluid compartment,[8–11] and it now seems that syrinx formation results from altered fluid balance between the SAS and ECS.[9] As only 1 small 10 mm neoplastic nodule induced extensive syrinx in our case, and based on previous reports of a higher protein concentration in neoplasm-associated syrinx and of syrinx cavity shrinkage after tumor removal,[3,9,11] transudation is likely to be the primary trigger for syrinx formation. Increased ECS fluid volume, viscosity and concentration, due to transudation from the neoplastic nodule, cause ECS flow to exceed capacity and obstruction of ECS flow towards the SAS. Thus, fluid accumulation in the central canal ensues. Moreover, by mass effect and syrinx formation, focal enlargement of the spinal cord adjacent to the neoplastic nodule might contribute to disrupted cerebral spinal fluid (CSF) flow and further extension of syrinx cavities associated with neoplastic nodules. CSF pulsatile-flow velocity is higher in the cervical area and then decreases towards the lumbar area.[9,11] Conversely, bulk flow and exchange of small-molecular-weight substances between the ECS and SAS are faster in areas of higher CSF flow velocity.[9] This corresponds with the finding that more syrinx occurs above the tumor than below it.[11] Patency of the central canal is also considered important in the development of non-communicating syringomyelia.[8]

In an autopsy study of 232 patients without spinal cord abnormalities,[12] varying degrees of central canal stenosis were present at 1 or more levels in 3% of infants aged < 1 year of age, 88% of adolescents and young adults, and 100% of individuals aged > 65 years. During the development of syringomyelia, central canal stenosis may limit the extent and continuity of central canal dilation and cause edematous change in the cord parenchyma.[8] This provides excellent explanations for the presence of multiple internal septa in the extensive syrinx cavity, and for edematous change in the cord parenchyma at level T12, in our patient.

Diagnostic features of spinal angiography for hemangioblastoma include enlarged feeding arteries, intense nodular tumor stains and early drainage veins.[3] Before the advent and popularity of whole-spine MRI, spinal angiography was considered the best and most accurate way to delineate tumor nodules, and there are still some indications for spinal angiography as part of the preoperative evaluation. For example, if the lesion is located in the lower thoracic or ventral aspect of the cord, it is important for the surgeon to demonstrate the relationship among the feeding arteries, the artery of Adamkiewicz, and the anterior spinal artery.[6,7] For surgical planning of one large hypervascular tumor with tortuous engorged tumor vessels, it is mandatory to evaluate the three-dimensional relationship between the tumor parenchyma and its feeding and drainage vessels in preoperative work-up.[6,7] It is probable that early coagulation of the main drainage vein before complete devascularization of all feeding arteries may change the hydrodynamics and contribute to surgical difficulties and spinal cord damage. Due to the high success rate of microsurgical techniques, and the potential complication of spinal cord infarction during endovascular embolization, embolization therapy may only be indicated when major surgery is contraindicated and when feeding arteries arise from the posterior spinal arteries. In some cases, magnetic resonance angiography of the spine can provide a noninvasive and effective technique for demonstrating intraspinal hypervascular lesions and engorged tumor vessels.[13]

Radiologic findings have been helpful in the diagnosis of spinal hemangioblastoma and differential diagnosis of astrocytoma, ependymoma, metastatic renal cell carcinoma in vHLD, paraganglioma, vascu-

lar anomalies, and other syringomyelia-associated conditions.[1,14–16] The finding of 1 well-demarcated, superficially located, intramedullary, intensely enhancing nodule with extensive syrinx formation is most likely hemangioblastoma.[2–4] Pre-operative recognition of spinal hemangioblastoma can limit the extent of surgery and alter the treatment strategy in vHLD.

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