Hemangioblastoma of the Spinal Cord Associated with Holocord Syringomyelia and Syringobulbia

Chain-Fa Su, Jin-Cherng Chen[1], Yung-Hsiang Hsu[2], Hung-Jen Yang, Chau-Chin Lee[3]

Department of Neurosurgery, Pathology[2], Radiology[3], Buddhist Tzu Chi General Hospital, Hualien, Taiwan; Department of Neurosurgery[1], Buddhist Dalin Tzu Chi General Hospital, Chia Yi, Taiwan

ABSTRACT

Hemangioblastoma occurring in the spinal cord is an uncommon but well-recognized entity. The present report shows a hemangioblastoma in the low thoracic cord uniquely associated with extensive rostral and caudal cysts to form a holocord syringomyelia and syringobulbia. This 36-year-old woman presented with progressive weakness and numbness over her right leg for about 3 years. She had difficulty walking for the previous two months. A neurological examination revealed spastic paresis of her right leg about grade 4 in power. Light touch, vibration and pin-prick pain sensations below thoracic dermatome level T11 were markedly reduced especially in the right lower limb. Gadolinium-enhanced magnetic resonance image (MRI) study demonstrated a 2 cm × 1 cm well-circumscribed mass at T11 in the spinal cord. Syringomyelia and cystic formation were noted rostral and caudal to this mass lesion. The upper syrinx extended from T11 up to the high cervical cord and to the lower medulla causing enlargement of the spinal cord. However, the lower one formed a cyst in the conus medullaris and the region of the cauda equina. During surgery to remove the masses, the big drainage vein was kept patent until the tumor was freed from the spinal cord. Internal debulking of the solid tumor was avoided and the tumor was taken out by a so-called "en bloc" pattern. Rostral and caudal cystic fluids drained spontaneously after tumor removal without shunting the syrinx to the subarachnoidal space. Post-operative MRI revealed total excision of the mass with a remarkable shrinkage of holocord cysts. Pathology showed a hemangioblastoma. Post-operative neurological function including urination and defecation did not change compared with that before surgery. (***Tzu Chi Med J*** 2001; **13**:187-192)

Key words: hemangioblastoma, spinal cord tumor, syringomyelia

INTRODUCTION

Hemangioblastomas are benign tumors of the central nervous system, comprising a dense network of capillaries or cavernous vessels. They occur most often at the posterior fossa with cystic formation. This tumor is less often encountered in the spinal cord. Early reports estimated the incidence of hemangioblastoma in the cord region to be about 1.6% to 3.3% of all intramedullary tumors [1,2]. However, a recent report revealed an incidence of 5.8% to 14.8% [3,4]. This increase of incidence might be attributed to the use of MRI examination of the spinal cord which can show intramedullary nodules, enlarged veins and an extensive syrinx well. Although spinal astrocytoma or ependymoma may be associated with a syrinx, hemangioblastomas growing in the cord usually demonstrate more uniform contrast enhancement and more extensive syrix formation [5]. In this presentation, we emphasize the significance of MRI study of a spinal cord hemangioblastoma and discuss the pitfalls of removing a hemangioblastoma in the spinal cord.

CASE REPORT

A 36-year-old woman presented with progressive

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Address reprint requests and correspondence to: Dr. Chain-Fa Su, Department of Neurosurgery, Buddhist Tzu Chi General Hospital, 707, Section 3, Chung Yang Road, Hualien, Taiwan

weakness in her right lower leg and increasing numbness over the anterior aspect of the right lower leg for about 3 years. She was seen by a Chinese herb doctor about 6 months before this admission and at that time, manipulation of her back caused severe back pain with radiation to the bilateral lower extremities. Because right lower limb weakness worsened over the past two months, she came to see us for further management. Past medical history was not contributory and there was no family history of this disorder. She had no complaints of upper limb dysfunction or sexual or urinary disturbance.

Physical examination revealed weakness of the thigh musculature and an evident weakness of dorsiflextion in her right big toe. Hypalgesia was noted over dermatomes, T11-S3 and light touch and thermal sensations were markedly decreased from L4 to S1 on the right. There was an impairment of vibration and position sense in the right lower foot. The patient had a wide based gait and she dragged her right foot when walking. Mental status, cranial nerves, and cerebellar function test were within normal limits. Eye fundus examination was normal.

Plain X-ray did not show significant abnormalities of the spine. However MR gadolinium-enhanced T1-weighted images showed a well-defined mass (Fig. 1. A, B) about 2 cm × 1 cm in size in the spinal cord. It was located at the T11 vertebral level and had a clear margin to the regional parenchyma. A rostral cystic formation could be identified at the thoracic and cervical cord. It extended up to the lower medulla to form a synringobulbia. In the cervical and thoracic cord, multiple horizontal septations transversing the syrinx cavity were prominent. (Fig. 2. A, B, C). The expanding cysts caused enlargement of the spinal cord. In addition, another cystic formation was found caudally to the tumor mass, forming a cyst at the conus medullaris and the cauda equina (Fig. 1, 2).

The patient received a total excision of the solid tumor. The walls of the rostral and caudal syrinx were not disturbed during the operation. After removal of the tumor mass, the cyst was opened and cystic fluid drained spontaneously. Briefly, the procedures are described as follows: (1) The pia and degenerated spinal cord tissue,

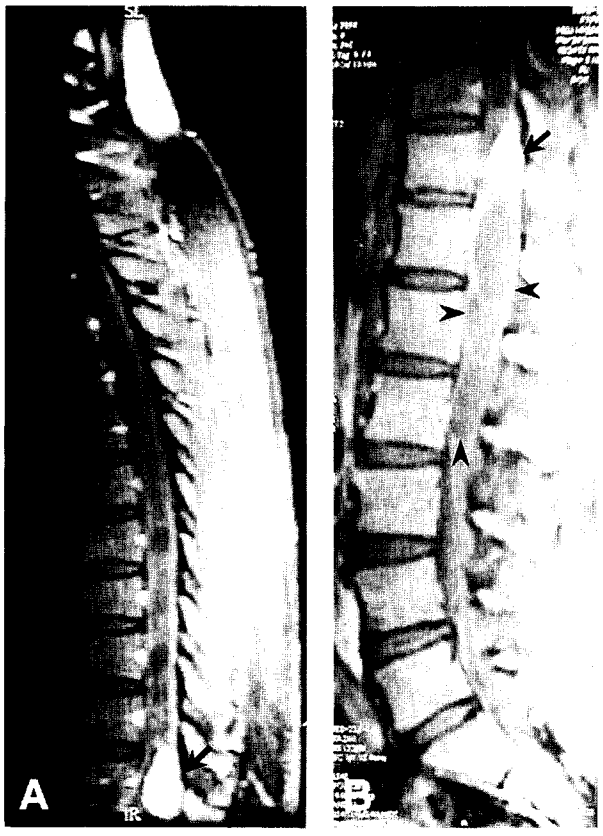

Fig. 1. Sagittal T1-weighted contrast-enhanced MR images. (A)preoperative image of thoracic region shows enhanced tumor (arrow) at the T11 level with rostral syringomyelia involving a long segment of the thoracic spinal cord. (B)Preoperative image of the lumbosacral region shows enhanced tumor and an enlarged cyst at the conus medullaris (small arrowheads).

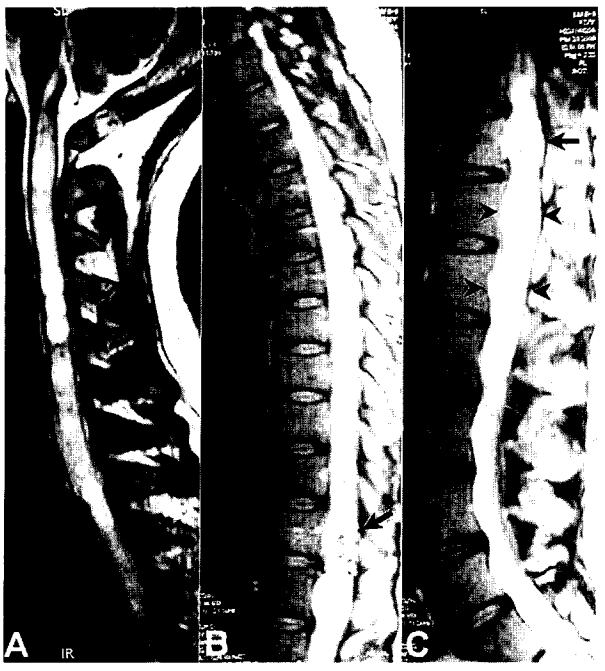

Fig. 2. Preoperative sagittal T2-weighted MR images. (A) MR image of the cervical region, (B)MR image of the thoracic region and (C)MR image of the lumbosacral region show extensive syringomyelia and mild syringobulbia. The tumor nodule at the T11 level (arrow) is mixed density in T2-weighted MR images.

which were compressed by tumor, were incised in an avascular area adjacent to the tumor located in the dorsal or lateral part of the cord. (2) The tumor dissection was carried out along the cleavage plane of the spinal cord. We made possible efforts to identify the interface between the tumor and the spinal cord. We also aspirated the tumor and the reactive glial tissue by using a small suction tip. By lightly pressing the spinal cord under the tumor with a small cottonoid and gently retracting the tumor with a bipolar forceps, the tumor could be dissected. (3) Internal debulking of the solid tumor was avoided and the drainage vein was well protected until the tumor was completely freed from the spinal cord. Pathological finding were consistent with a hemangioblastoma (Fig. 5). A follow-up MR examination was performed 6 weeks later following surgery and it showed a significant reduction in the size of the holocord syrinx. Only thin collapsed syrinx signal could be visualized (Fig. 3. B). MRI showed total removal of this tumor and the cord tissue became thin after surgery (Fig. 4). The patient had no neurological function deterioration after surgery and she was able to return to work.

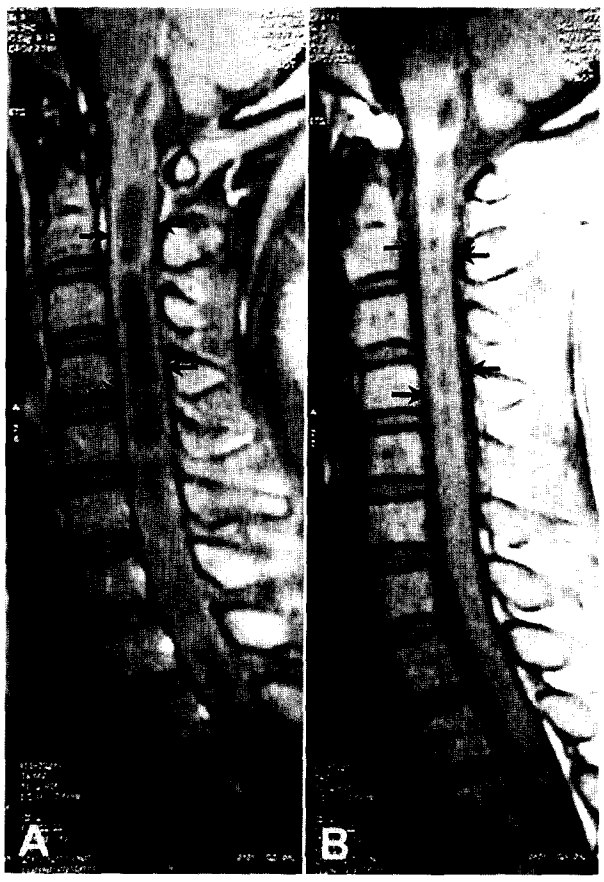

Fig. 3. T1-weighted sagittal MR images of the cervical spinal cord. (A)Preoperative MR shows extensive syrinx with septations at the cervical cord (arrows) and lower part of the medulla. (B)Postoperative MRI shows collapse of the syrinx (arrows).

DISCUSSION

Hemangioblastoma as a spinal cord tumor is rare

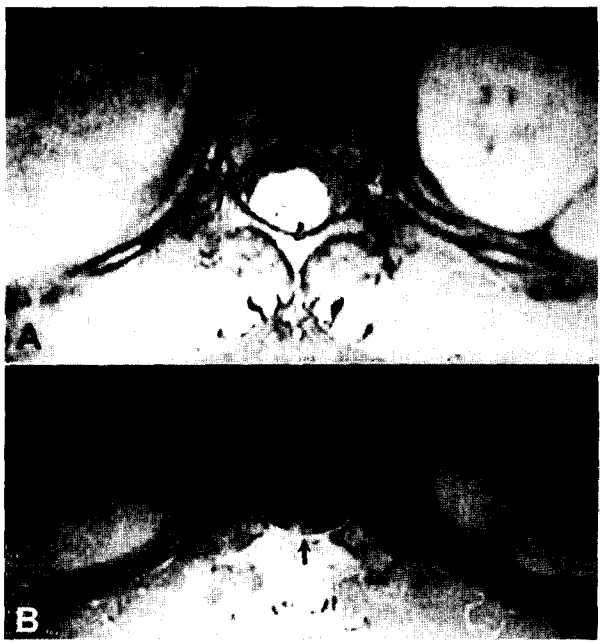

Fig. 4. Gadolinium-enhanced axial MR images of the spinal cord at T11. (A)Preoperative MRI shows an enhanced tumor mass occupies the dorsal part of the cord (large arrow). (B)The tumor was totally removed with a small part of cord tissue left (between 2 small arrows).

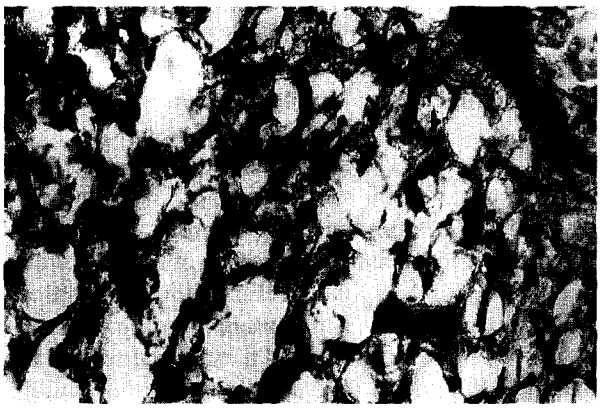

Fig. 5. Microscopic appearance of the hemangioblastoma. Numerous capillary channels lined by a single layer of endothelial cells form a plexiform pattern. (Hematoxylin and eosin stain, × 400)

but recent reports show an increase in its incidence because of MRI studies. Previously, hemangioblastomas with cyst formation in the cord could have been misdiagnosed as astrocytomas because clinicians lacked definite pathological proof. Complete MRI examination can help diagnose a hamangioblastoma by demonstrating the clear margin of the cord tumor mass with the surrounding cyst. This enables the neurosurgeon to reduce the invasiveness and risks of surgical excision [7,4].

Approximately 66% of spinal cord hemangioblastomas are associated with syringomyelia [1,8-10]. In our patient, we demonstrated a unique extension of the cysts to form a holocord syringomyelia and a syringobulbia. There are several theories about the pathogenesis of syringomyelia associated with the spinal cord tumors. Barnett and Rewcastle [11] stated that syrinx associated with tumors are secondary to CSF flow obstruction during periods of raised intraspinal pressure. Other authors however favored the theory that in cases of hemangioblastomatous syringomyelia, the tumor transudes fluid that dissects through the gray matter alongside the central canal [1,9]. Therefore, gliosis is commonly associated with syringomyelia [12].

Surgery is considered to be an effective therapy for most benign tumors of the spinal cord including hemangioblastoma [13,14]. To remove a hemangioblastoma of the spinal cord is relatively difficult, because the cord diameter is small and it has extensive functional nervous tissue. In addition, quadriplegia or paraplegia can occur even with microsurgical techniques. Because intramedullary hemangioblastoma of the spinal cord is generally insensitive to radiotherapy, radiotherapy is not a recommended treatment, although some reports of symptomatic improvement have been addressed [15,16].

There are several pitfalls to avoid when removing a hemangioblastoma of cord [4,6,17,18]. Herrmain et al emphasized the use of CO_2 laser [6,19] but Brotchi et al stressed the use of meticulous bipolar coagulation and following the rules of "en bloc" excision, to avoid removing the tumor piece by piece. Ultrasonic aspirator debulking is also contraindicated [20]. Xu reported microsurgical treatment of an intramedullary hemangioblastoma of the spinal cord in 13 cases and strongly stressed that coagulation and division of the main draining vein should be avoided before complete devascularization. A change in hemodynamics may increase the difficulty of the procedure and damage the spinal cord itself [4]. We preferred to separate, coagulate and divide the feeding arteries in the dorsal and lateral areas initially. Then we approached the end of the tumor to avoid damaging the main draining vein. The syrinx was entered, and fluid drained spontaneously. In this particular case we proved that extensive cysts in the whole cord could diminish after the tumor was removed and syringo-subarachnoidal shunting was unnecessary. The syringes were dramatically reduced or disappeared uniformly.

In conclusion, MRI study is the most effective method to evaluate spinal cord hemangioblastoma. Radiotherapy to treat a benign lesion of the cord such as a hemangioblastoma is not recommended. Surgical removal of the tumor mass without disturbance of the syrinx wall is the basic strategy but it should follow principles such as protecting the drainage vein and avoiding central debulking of the tumor mass.

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脊髓血管母細胞瘤合併全脊髓空洞症－病例報告

蘇泉發　陳金城[1]　許永祥[2]　楊宏仁　李超群[3]

佛教慈濟綜合醫院神經外科　病理科[2]　放射線科[3]　佛教大林慈濟綜合醫院神經外科[1]

摘要

脊髓血管母細胞瘤在脊髓腫瘤中本已少見，而本文報導一例胸髓之脊髓血管母細胞瘤合併全脊髓空洞之上下延伸，更屬罕見。患者是一位36歲女性，自3年前即逐漸發生右下腳無力並合併麻感，入院前2個月；右下腳無力更形明顯，而致行走困難，本院門診時，理學檢查顯示右大腿及下腳無力約4級肌力，自第11胸椎皮節以下疼痛感及輕觸感均明顯下降，且振動及體位感覺在右下腳顯示缺損。磁共振造影，經對比劑注射顯影，顯示第11胸髓有約2×1公分大之脊髓內腫瘤。腫瘤與正常組織邊緣清楚且大部份位於脊髓神經之背部並造成脊髓神經之壓迫。沿著腫瘤向上有一連續之脊髓空洞症，由胸髓沿伸至頸髓，並延至腦幹處，形成長形之全脊髓空洞症，而本身向下更在脊髓圓錐部至第一、二腰椎間形成囊腫。手術採顯微手術沿著腫瘤邊緣，將腫瘤完全取出，但對於頸、胸髓之脊髓空洞症及圓錐部之囊腫則並未刻意引流或行導管手術，術後六週，經行磁共振造影檢查，顯示胸椎第11節脊髓之腫瘤已切除乾淨，且頸部及胸髓之脊髓空洞症也明顯萎縮，患者之神經功能在術後，並未變壞，目前僅呈右腳輕度強直性無力，且排尿、排便功能正常，病理顯示為血管母細胞瘤。(**慈濟醫學** 2001; **13**:187-192)

關鍵語：血管母細胞瘤，脊髓腫瘤，脊髓空洞症

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