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Spinal Cord Hamartoma
- A Case Report -

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Spinal cord hamartoma is rare disease. Many authors reported that most of hamartomas occured in patients with tuberous sclerosis, spinal dysraphism or neurofibromatosis type I. There were very few cases unrelated with these disease throughout the world.

We present a case of spinal cord hamartoma which is not associated with tuberous sclerosis or neurofibromatosis and describe the clinical presentation, radiological appearance, pathological features and treatment of this unique disease entity.

A 29-year-old female patient presented with progressive myelopathy for 7 months. Magnetic resonance imaging revealed a thoracic intramedullary tumor that was extended from T1 to T4. The tumor was resected through the posterior approach and the patient improved postoperatively.

We report a rare case of spinal cord hamartoma that was verified pathologically. Although the magnetic resonance imaging may be helpful in determining the extent of this lesion, it cannot be used to distinguish neoplastic lesions from non-neoplastic ones. Microsurgical resection is considered to the therapeutic option for these disease.

Key Words: Hamartoma · Thoracic · Spinal cord tumor

INTRODUCTION

Hamartomas are not tumor but benign, tumor-like proliferation composed of normal mature cells and tissues normally present in the affected part. Central nervous system hamartomas rarely occur and are usually supratentorial lesions combined with tuberous sclerosis or hypothalamic tumor[8,13]. In infratentorium, they are usually associated with either spinal dysraphism[6,16] or neurofibromatosis (NF) Type 1[2,7,9]. Spinal cord hamartomas not related with these diseases are extremely rare. We present a case of intramedullary spinal cord hamartoma and discuss the clinical presentation, radiological appearance, pathological features and treatment.

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CASE REPORT

A 29-year-old female admitted to our hospital in May, 2003. She complained of radiating pain and weakness of both legs predominantly on her left side. The leg pain were mainly S1 dermatome area and bladder or bowel dysfunction was not noted. She had noticed these symptoms 7 months before the admission, and the symptoms slowly progressed. She had no previous medical illness.

1. Physical examination

A physical examination revealed hypesthesia on light touch and pain below T4 dermatome area. The examination also revealed decreased muscle strength of both lower extremities, especially

in left side; Hip (G4/4), knee G4+/4-), ankle (G4+/4+), big toe (G4+/3). But there were no wasting of muscle. Deep tendon reflexes were increased on the both lower extremities. Bilateral ankle clonus were noted and Babinski's sign were also positive. Other examinations showed no abnormality.

2. Imaging

Thoracic plain radiographs showed no gross bony abnormality. Computerized tomography taken at private hospital showed unusual bulbous dilated upper thoracic spinal cord with low density suggesting an intramedullary tumor with lipomatous component. Sagittal T2-weighted magnetic resonance (MR) imaging of C- and T-spine demonstrated an intramedullary and extramedullary combined mass in T1-T4 level. T1-weighted image showed a bright signal intensity at posterior aspect of spinal cord. After Gd-DTPA enhancement, other abnormalities were not vis- ualized in the spinal cord(Fig. 1). Microsurgical resection of the tumor was performed.

3. Surgical operation

The patient was placed in prone position and laminectomy was done from T1 to T4 using a high speed air drill. After the midline dural incision, there was no arachnoidal adhesion and the spinal cord had been expanded with intermingled mass. The outer surface of the mass was yellowish with fat component and had hard consistency. The mass was poorly demarcated from the normal spinal cord tissue and the dissection plane like tumor capsule was not discrete. A piece of tumor tissue was sent for frozen section and it proved to be consistent with fibroadipose tissue. Then, the mass was meticulously removed using a sharp hook on the basis of different consistency with normal spinal cord tissue. The tumor vascularity was not so high and the blood loss was minimal. After hemostasis, the dura was closed in a watertight fashion and the wound was closed in layer by layer technique.

4. Pathological finding

The specimen consisted of tiny pieces of gray white tissue. The light microscopic examination revealed spinal cord tissue mainly composed of many longitudinal neural tracts and blood vessels. The spinal cord tissue was intermingled with disorganizing fat tissue, nerve fibers, fibrocollagenous tissue(Fig. 2). The pathological findings were consistent with hamartomatous tissue.

5. Clinical course

After the operation, the both leg pain was improved and the decreased muscle strength was increased; Hip (G5/5), knee (G4+/4+), ankle (G4+/4+), big toe (G4+/4). The spasticity of the lower extremities was also markedly improved, but the hypesthesia of

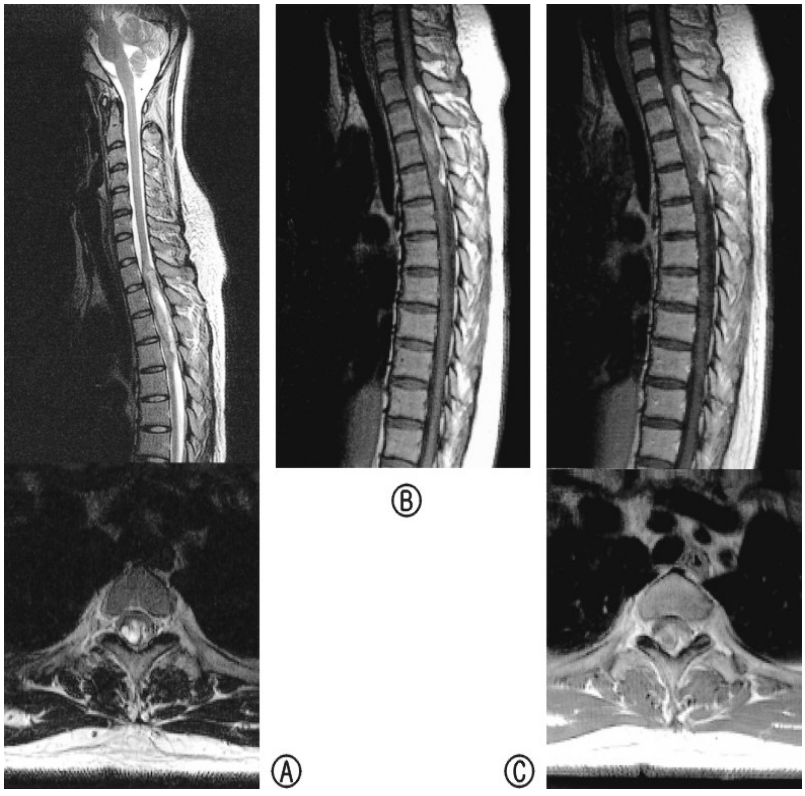

Fig. 1. Preoperative magnetic resonance imaging of thoracic spine. A; Sagittal(Up) and axial(Down) T2-weighted image showing intramedullary and extramedullary combined mass lesion in T1-T4. B; Sagittal T1-weighted image showing high signal intensity at posterior aspect of spinal cord and epidural space. C; Gd-DTPA enhanced sagittal(Up) and axial (Down) T1-weighted image showing no abnormal increased signal intensity.

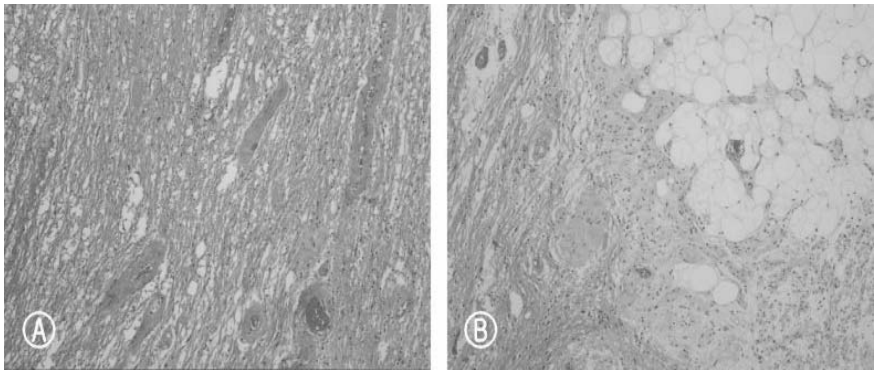

Fig. 2. Pathologic images. A; Spinal cord tissue composed of many longitudinal neural tracts and blood vessels(H-E stain, ×100). B; Nervous tissue intermingled with fat tissue and fibrocollagenous tissue(H-E stain, ×100).

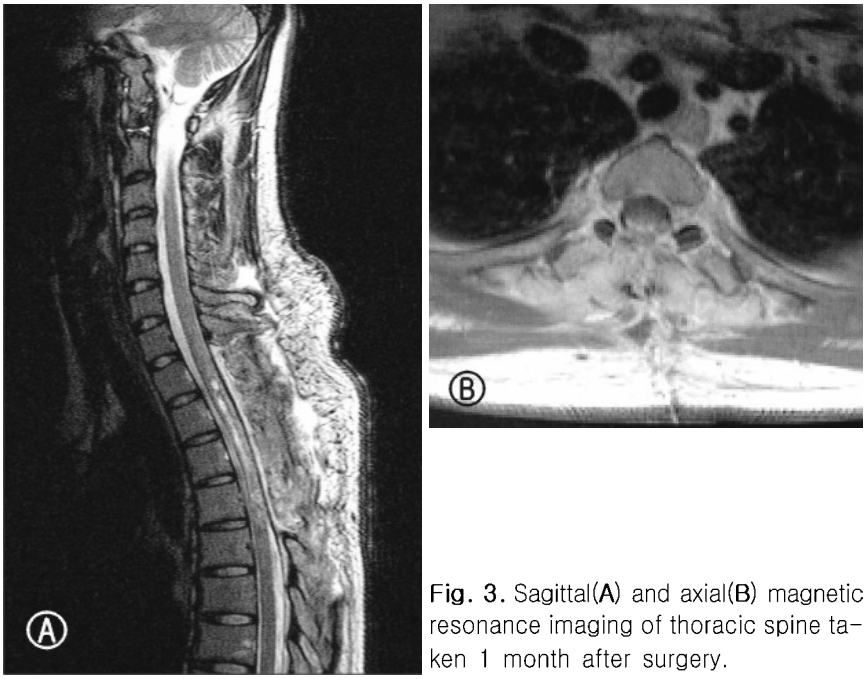

Fig. 3. Sagittal(A) and axial(B) magnetic resonance imaging of thoracic spine taken 1 month after surgery.

both lower extremities was slightly improved. The patient was discharged on postoperative day 16. At follow up 15 months after operation, the motor weakness of both lower extremities were fully recoverd and only minimal hypethesia of left lower extremity was remained. The follow up MR imaging demonstrated no residual tumor(Fig. 3, 4).

DISCUSSION

The spinal cord hamartomas are very rare disease entity. Several authors described central nervous system hamartomas are frequently occurred in patients with tuberous sclerosis[13]. However, spinal cord hamartoma not associated with neurofibromatosis (NF) type 1 and spinal dysraphism is very rare. There were only two reported cases. One was reported by Riley et al[11] in abroad, and the other by Lee et al[17] in domestic.

Spinal hamartomas are usually located in the thoracic, lumbar, and sacral regions or at the junctions between these segments. Morris et al[10] announced that nearly half of the hamartomas were located in the thoracolumbar junction. In our case, it was located in upper thoracic region.

Spinal cord hamartoma related to the NF are usually asymptomatic[10,15]. These lesions are benign and tend to decrease in size as the patients get older[1,12]. Many authors reported that spinal hamartomas neither grow excessively nor show malignancy, and the prognoses are good[3,10].

The pathogenesis for formation of spinal hamartoma is not exactly known. Some authors suggested incomplete or premature disjunction during the neural tube formation is possible mechanism[3]. Hamartomas associated with NF type 1 consist primarily of neural crest-derived neuron, glial cell, schwann cell, fibroblast, vascular endothelial cell and mast cell[5,8]. Brownlee et al[2] reported that they were composed of proliferations of glial cells, ganglion cells, disoriented axons, and vessels. They have termed it vascular and neuroglial hamartomas. On the other side, so-called midline spinal hamartomas may present with a skin dimple, cutaneous angioma, subcutaneous mass, or normal overlying skin[10,15]. Particularly, midline spinal hamartomas associated with the spinal dysraphism are often mistaken as meningomyelocele, lipomeningomyelocele, myelocystocele, meningocele, spinal lipoma, and teratoma. However, it is not connected with the neurological

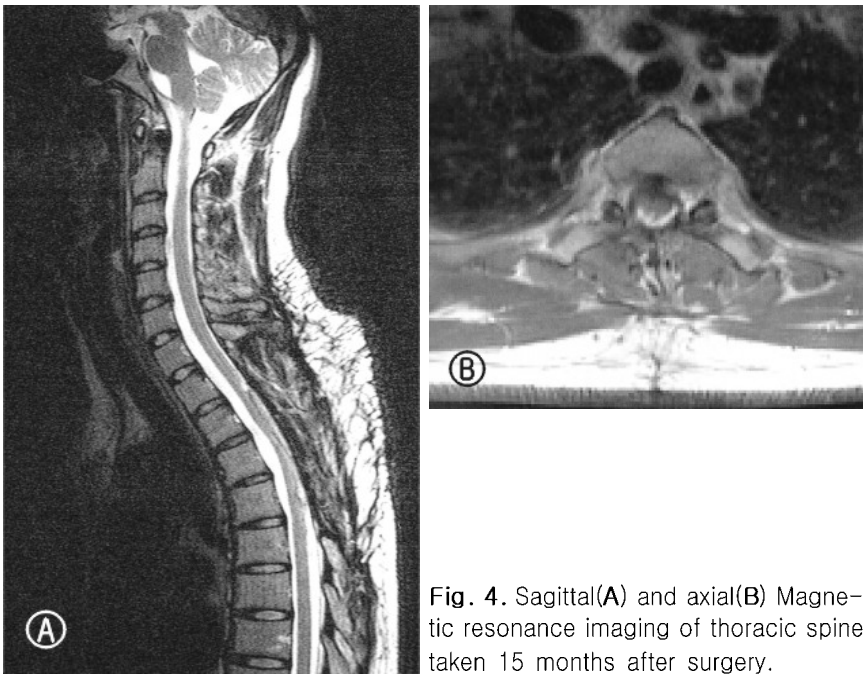

Fig. 4. Sagittal(A) and axial(B) Magnetic resonance imaging of thoracic spine taken 15 months after surgery.

complication, and musculoskeletal abnormality or scoliosis[10]. In our case, it is applicable to neuroglial hamartoma because pathological finding revealed the intermingled fat tissue, nerve fibers, fibrocollagenous tissue.

Spinal hamartomas differently diagnosed from other masses such as teratoma, epidermoid, dermoid. They may be especially difficult to distinguish them from teratomas, which arise from displaced germ cell. Teratoma are distinguished from harmartomas only by pathological examination. Teratomas are composed of elements from three immature germinal layers. On the contrary, spinal hamartomas are composed of only ectodermal and mesodermal, mature and well-differentiated elements, They do not contain elements from all three germ cell layers, as teratomas do. The teratoma is more tumorous condition than other intramedullary tumors such as lipomas or hemangiomas[10,15].

In order to distinguish hamartoma from other masses, many authors have reported that MR imaing is useful modality but its finding is variable case by case.

Castillo M et al.[3] suggested that hamartomas appeared isointense with the spinal cord on all MR imaging sequences. Riley et al.[11] presented slightly hyperintense on the T1-weighted MR image and isointense on the T2-weighted MR image and no contrast enhancement. In our case, T1 and T2-weighted image showed mixed high signal intensity. Although the myelographic and computed tomographic appearance of these lesions were similar, MR imaging was proved to be useful in establishing and excluding the other differential diagnosis[2,4,14]. Also, MR imaging clearly demonstrated the intramedullary, exophytic nature of the lesion. Furthermore, the lack of contrast enhancement essentially excluded many of the lesions in the broader differential diagnosis. The inhomogenous contrast enhancement in MR imaging is a distinctive characteristic of teratomas. While epidermoids have a signal intensity similar to fluid on both T1 and T2-weighted images, der- moids have a bright signal intensity on T1-weighted images.

The treatment options of spinal hamartoma are various from non-operation to surgical excision and detethering of arachnoid membrane. Morris et al[10] described that surgical operation is usually performed to prevent infection or neurologic damage resulting from tethering of the cord, and it is also used to correct cosmetic defect.

But, other authors presented five patients with postoperative cervical spinal cord tethering who improved after surgery for detethering[11]. They insisted the neurological symptoms in the patient of spinal hamartoma are due to spinal cord tethering form arachnoid adhesion overlying it. If neurological abnormality is not found, the operation of spinal cord hamartoma is not logical. However, when neurological abnormality was found as our case, the removal of spinal cord mass could be the proper treatment.

CONCLUSION

Spinal cord hamartoma is a very rare disease entity. Patients are usually asymptomatic or have only minimal neurologic deficits. MR images is very helpful for diagnosis but is not confirming tool. For patients with neurological abnormality, surgical removal of mass is proper as diagnostic, pathological confirma-

tion and treatment.

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