

Case Report

Intramedullary teratoma of the spine: report of two adult cases

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Study design: Two case reports of intramedullary teratoma in the spinal cord of adults, and a review of the literature.

Objective: To investigate and describe unusual cases of spinal teratoma using MRI to define features that may be used to avoid misdiagnosis.

Setting: A department of orthopedic surgery in Japan.

Methods: One patient, a 37-year-old woman, was referred because of gait disturbance. She was evaluated by myelography, CT scan with myelography, and MRI. T12 through L1 laminoplasty was performed and the tumor was subtotally removed. The other patient, a 56-year-old man, was referred because of muscle weakness and sensory disturbance. MRI revealed multiple spinal tumors. C4 through C6 laminoplasty and T12 through L2 laminoplasty were performed, and the tumors in these regions were subtotally removed.

Results: In Case 1, the postoperative course was excellent, and histological examination of the resected specimen revealed a spinal teratoma consisting of ectodermal and mesodermal elements. In Case 2, the symptoms were resolved after surgery, and ectodermal, mesodermal and endodermal elements were revealed.

Conclusions: Although intramedullary teratomas are very rare in adults, they need to be considered in differential diagnosis.

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Introduction

Intramedullary teratomas in the spinal cord are very rare, especially in adults over 50 years of age. We present two adult patients with spinal teratoma, and discuss the diagnosis of the condition and its surgical treatment.

Case reports

Case 1

A 33-year-old woman presented at the outpatient center of the Department of Orthopaedic Surgery, Gifu University School of Medicine, in August, 1987, with a 1-year history of gait disturbance. She had difficulty in walking because of muscle weakness in her lower extremity, but she was able to walk without assistance.

She had also noticed numbness in her feet for at least 5 years. MRI of the thoracic and lumbar spine demonstrated a spinal cord tumor at the T12 and L1

levels. By August 1991, her gait disturbance had become severe, so she was admitted to our hospital. On the day of admission, the results of general physical examination were unremarkable. However, neurological examination revealed bilateral paresis of the lower leg and foot, and bilateral lower-extremity hyperreflexia with ankle clonus. The plantar reflexes were flexor. Pain sensation and thermo sensation were markedly decreased bilaterally below the L4-level. Examination of the cerebrospinal fluid revealed a protein concentration of 715 mg/dl.

A plain X-ray film of the spine showed marked widening of the interpedicular space at the T12-L1 levels with erosion of the left pedicle and L5 spina bifida. MRI demonstrated a heterogeneous mass extending from the T12 to L1 level (Figure 1). An Iotrolan myelogram demonstrated a complete extradural block at the upper margin of L2. Iotrolan CT-myelography demonstrated no Iotrolan ring at the conus medullaris (T12-L1) because of the tumor. At the L2 level, the distal tip of the conus and cauda equina was markedly compressed, with leftward and ventral shift.

On September 2, 1991, reconstructive laminoplasty of T12 through L1 with tumor resection was

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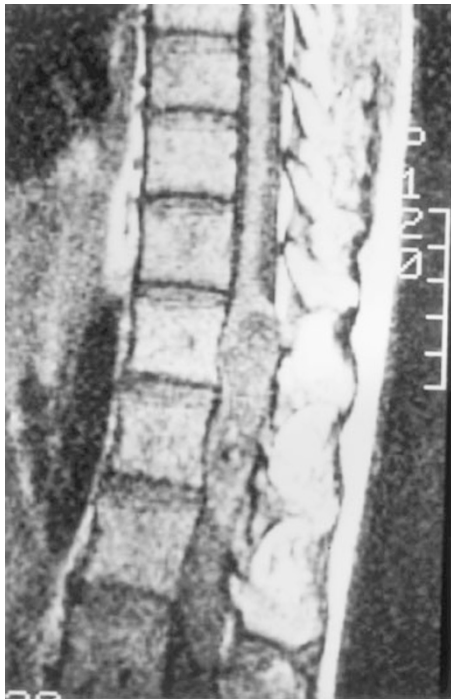


Figure 1 Sagittal MR image (T1-weighted) of Case 1, showing a heterogeneous intracanalicular mass extending from T12 to L1

performed. When the dura mater was opened, a pinkish-yellow tumor was found. The tumor extended between two vertebral levels (T12-L1). Inside the tumor capsule, a yellowish fatty mucoid substance containing fragments of bone and hair were found. The tumor was found to be intramedullary in origin. After careful aspiration of the mucoid substance, the capsule of the tumor was subtotally resected. A small portion of the cauda equina was excised because of its firm adhesion to the tumor capsule.

Histological examination showed that the capsule was composed of stratified squamous epithelium with hair and mucous glands. Inside the capsule there was mature bone tissue surrounded by connective tissue. Histological analysis demonstrated a mature teratoma with two components: ectodermal elements and mesodermal elements.

Four weeks after the operation, the patient was able to walk with a solid trunk brace. Eight years after the operation, she showed considerable neurological improvement, and no sign of tumor recurrence in an MRI study.

Case 2

A 56-year-old man came to our hospital in March 1996 with a 2-year history of muscle weakness in the right lower leg. He had also noticed sensory disturbance in all four limbs. MRI of the spine revealed multiple spinal cord tumors. Around April 1996, his gait

disturbance gradually became severe, and he was admitted to our hospital. On the day of admission, the results of general physical examination were unremarkable. However, neurological examination demonstrated bilateral patellar tendon hyperreflexia. The plantar reflexes were flexor. Touch sensation was markedly decreased bilaterally below the T10 level. There was also muscle weakness in the lower legs, necessitating use of a cane for walking.

A plain X-ray film of the spine showed no abnormal signs. MRI demonstrated heterogeneous masses extending from the medulla oblongata to the L2 level (Figure 2a,b). Iotrolan myelography demonstrated complete extradural block at the L2 level. Iotrolan CT-myelography failed to demonstrate an Iotrolan ring at the conus medullaris (T12-L1) because of the tumor. At the L2 level, the distal tip of the conus and cauda equina was markedly compressed, with leftward and ventral shift.

On May 20, 1996, laminoplasty of T12 through L2 and of C4 through C6 was performed, with resection of the tumors in both regions. When the dura mater in the thoracolumbar region was opened, lipid flowed out. The border between the tumor and conus medullaris was indistinct. We therefore removed the tumor within a safety margin. When the dura mater in the cervical region was opened, lipid flowed out but there was no mass.

Histological examination showed that the capsule was composed of stratified squamous epithelium. Inside the capsule, there were mucous glands, bone tissue, smooth muscle fibers, and peripheral nerve bundles surrounded by dense hyalinized tissue with calcification. Histological analysis demonstrated a mature teratoma with three components: ectodermal, mesodermal and endodermal elements.

Eight days after the operation, the patient was able to walk with a solid trunk brace, a Philadelphia collar, and a cane. At present, he suffers from urinary disturbance and paralysis, making it difficult for him to negotiate stairs and perform fine finger movements.

Discussion

Although sacrococcygeal teratoma in newborn babies has frequently been reported, spinal teratoma in adults is very rare. Teratomas accounts for only 0.2–0.5% of all spinal cord tumors,^{1–4} and only 2% of all teratomas occur in the central nervous system. Furthermore, only a very small proportion of CNS teratomas occur in the spinal cord.^{5,6} Only seven such cases have been reported in patients aged over 50 years of age (Table 1).^{1,7–12} In adult cases of spinal teratoma, the thoracolumbar region is most commonly affected.⁸ However, multiple occurrence has not been reported before. Our present Case 2 is considered to be very rare because the patient was 56 years old, and the tumors extended from the medulla oblongata to the L2 level.

The origin of spinal cord teratoma is controversial, and three hypotheses have been proposed. One is that

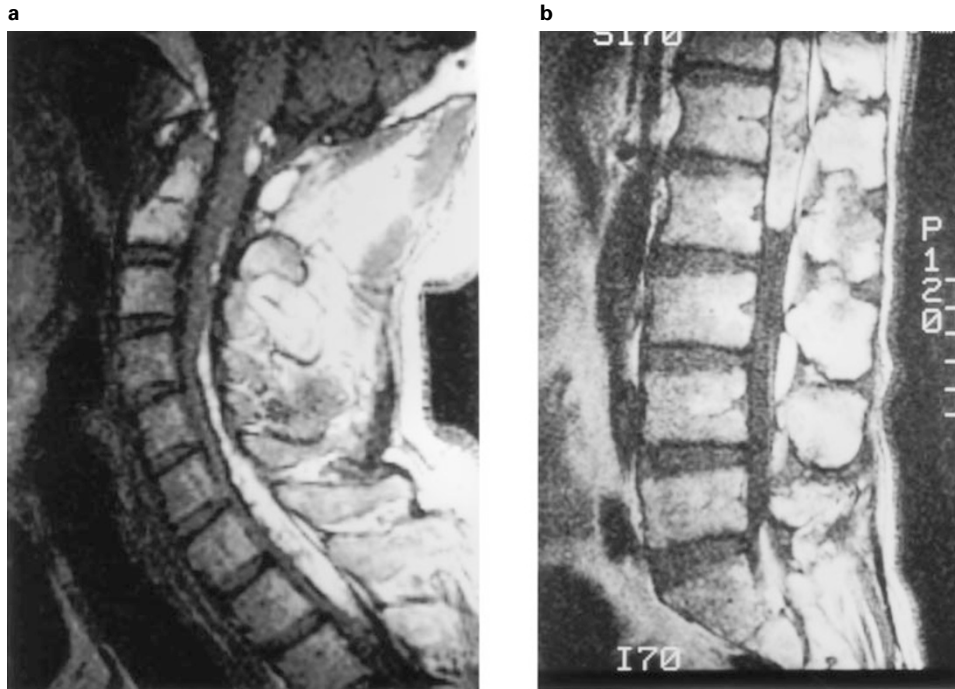


Figure 2 (a) Sagittal MR image (T1-weighted) of Case 2, showing a heterogeneous intracanalicular mass extending from the medulla oblongata to the T2 level. (b) Sagittal MR image (T1-weighted) of Case 2, showing a heterogeneous intracanalicular mass extending from T12 to the L2 level

Table 1 Reported cases of spinal teratoma in older patients

Author	Age (years)	Sex	Location
Bakay (1956) ⁹	65	F	L1-L2
Sloof (1964) ¹	67	M	S
Nakayama (1938) ⁸	51	F	T12-L3
Monajati (1986) ¹⁰	52	M	L3-L5
Hamabuchi (1989) ⁷	54	M	T12-L1
Kaji (1993) ¹¹	79	F	T11-L2
Okuyama (2000) ¹²	50	F	L3-L4
Case 2 (1996)	56	M	Multiple

such teratomas represent abortive 'twin' formation.¹³ Another is that teratomatous cysts represent the ependymal diverticulum of the central canals of the spinal cord.¹⁴ However, this theory does not explain the presence of muscle, bone, fat and thyroid-like glandular tissue, which are not derived from the neuroepithelium.¹⁵ The third theory, which seems to be supported by the majority of authors, is that teratoma arises from multipotential germinal cells that are misplaced in early embryonic development.^{16,17} Rewcastle, however, has suggested that teratoma arises not from simple misplacement of normally developing somatic cells but from fusion of two misplaced haploid germ cells.¹⁸

Originally, tridermal tumors were termed teratomas.¹⁷ Although a teratoma is a true neoplasm composed of all three germinal layers, the presence of only two germinal components does not necessarily rule

out this diagnosis. Derivatives of one or two germ layers tend to overgrow the others, so that the total number of germ layers may be difficult to ascertain. In our present Case 1, the tumor was found to be bidermal, as has often been the case for teratomas reported previously.^{7,19,20}

Spinal teratomas do not show any specific subjective symptoms. Plain X-ray films often show erosion of vertebral bodies and widening of the interpedicular space, with or without associated vertebral anomalies at the same level, such as spina bifida,^{16,21-24} vertebral body fusion,⁹ asymmetry of the vertebral bodies,¹⁸ and diastematomyelia.²⁵ In Case 1, the tumor was located at the same level as the vertebral anomalies (fused laminae). CT is very useful for preoperative diagnosis because teratomas exhibit variable density or calcification.⁸ MR imaging is the most valuable diagnostic technique for teratoma, because the tumors exhibit mixed high and low intensity, indicative of tissue heterogeneity.²⁶ In the present cases, MRI revealed the local size of the tumor and variable signal intensity inside the tumor (probably representing bone, fat etc.).

Total resection is the treatment of choice, but this is almost impossible without some injury to the neural tissue. Intimal adhesion of the teratomatous cyst to the surrounding nervous parenchyma is seen in about 50% of cases.²⁰ However, subtotal removal with no subsequent recurrence of symptoms over many years due to the extremely slow tumor growth has been reported.^{18,20} It is important to resect as much of the tumor as possible while preserving all neural tissue.

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