

Case Report

Spinal Intradural Extramedullary Mature Cystic Teratoma in an Adult

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Spinal intradural extramedullary teratoma is a rare condition that develops more commonly in children than in adults and may be associated with spinal dysraphism. We report a rare case of adult-onset intradural extramedullary teratoma in the thoracolumbar spinal cord with no evidence of spinal dysraphism and without the history of prior spinal surgery. The patient was a 38-year-old male whose chief complaint was urinary incontinence. X-ray images of the thoracolumbar spine showed the widening of the interpedicular distance and posterior marginal erosion of the vertebral bodies and pedicles at the T11, T12, and L1 level. Magnetic resonance imagings of the lumbar spine showed a lobulated inhomogeneous high signal intradural mass (87 × 29 × 20 mm) between T11 and L1 and a high signal fluid collection at the T11 level. Laminectomy of the T11-L1 region was performed, and the mass was subtotaly excised. The resected tumor was histopathologically diagnosed as a mature cystic teratoma. The patient's symptom of urinary incontinence was improved following the surgery.

KEY WORDS : Spinal cord neoplasm · Mature teratoma.

INTRODUCTION

Teratomas involving the central nervous system are rare. The incidence of intracranial teratomas is low, and these tumors account for approximately 0.5-2.2% of all intracranial tumors⁴. Further, spinal teratomas account for approximately 0.1-0.5% of all spinal tumors^{5,9}. Slooth et al.¹ identified spinal teratomas in only 2 of 1,322 patients with primary spinal cord tumors (reviewed in 1964); this emphasizes the rarity of these tumors. Cybulski et al.⁴ conducted a literature review in 1984, and they found that only 56 cases of primary spinal teratomas had been reported at the time. In 1999, Poeze et al.¹⁰ reported that only 83 spinal teratoma cases had been published until then. We describe a rare case of a spinal intradural extramedullary mature cystic teratoma in an adult.

CASE REPORT

A 38-year-old male patient presented with back pain that

had persisted for approximately 8 years and urinary incontinence that had lasted approximately 2 months. He had neither history of spinal surgery nor any other spinal procedures. On physical examination, no abnormal hairy patch or dimple was noted on the back. Neurological examination revealed that the anal tone was within the normal range and that the bulbocavernosus and cremasteric reflexes were normal, despite the urinary incontinence. A laboratory study results were within normal ranges.

X-ray images of the thoracolumbar spine showed the widening of the interpedicular distance and posterior marginal erosion of the vertebral bodies and pedicles at the T11, T12, and L1 level (Fig. 1). Three-dimensional computed tomography (CT) revealed the presence of a mixed-density lesion that occupied the intradural space and widening of the spinal canal at the T11-L1 level (Fig. 2). Sagittal T2-weighted magnetic resonance imaging (MRI) of the lumbar spine (L-spine) revealed the presence of a lobulated inhomogeneous high signal intradural mass between T11 and L1 and a high signal fluid collection at the T11 level. The mass lesions extended from T11 to L1 and were in contact with the conus medullaris. None of the lesions were enhanced following gadolinium injection (Fig. 3).

The operation was performed in the prone position. The procedure involved total laminectomy at the T11-L1 levels.

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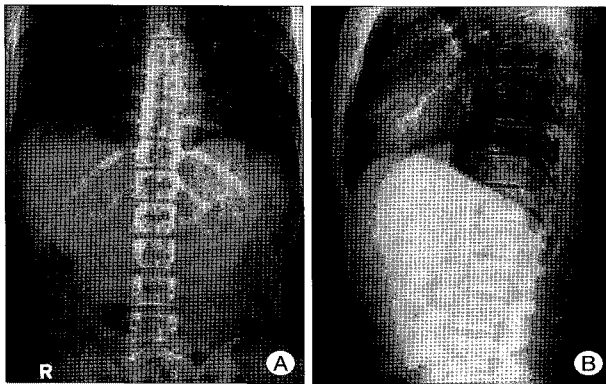


Fig. 1. X-ray images of the thoracolumbar spine (A and B) show the widening of the interpedicular distance and posterior marginal erosion of the vertebral bodies and pedicles at the T11, T12, and L1 level.



Fig. 2. Three-dimensional computed tomography revealing the presence of a mixed-density lesion that occupy the intradural space and widening of the spinal canal at the T11-L1 level.

The dura mater was opened via a mid-line incision to expose the tumor. A yellowish fatty cyst was found, the needle-aspiration was first done to prevent the leakage of cystic fluid into the intradural space. The hairy substance was found in the aspirated fluid, and the adjacent spinal cord was compressed by the cystic mass inferiorly. Subtotal tumor resection was performed under a surgical microscope. The distal fibrous solid mass was adhered strongly to the conus medullaris and the neural filament (Fig. 4). The excised mass contained 4 components : (1) lumps of grayish yellow cheese material containing hair follicles, (2) a fragment of whitish fibromembranous calcified tissue, (3) several fragments of gray soft tissue, and (4) a fragment of fibromembranous tissue.

Histopathological examination of the excised mass revealed the presence of elements of the 3 germ cell layers the ectoderm, mesoderm, and endoderm. Microscopic findings revealed that the cystic wall contained keratinous squamous epithelial cells and keratin lamellae, implying the ectodermal origin. Moreover, mature fat cells and bone components, indicating the mesodermal origin, were also found in

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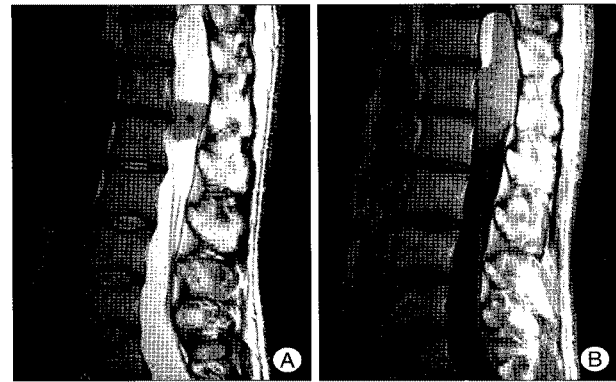


Fig. 3. Sagittal T2-weighted (A) and T1-weighted gadolinium enhanced (B) magnetic resonance imaging of the lumbar spine (L-spine) showing the presence of a lobulated inhomogeneous high signal intradural mass between T11 and L1 and a high signal fluid collection at the T11 level. The mass lesions extend from T11 to L1 and are in contact with the conus medullaris. None of the lesions are enhanced following gadolinium injection.

distal solid mass. Furthermore, the enteric glandular cells and respiratory epithelial cells of the endodermal origin were noted in mucinous cystic mass. The final diagnosis of the mass was a mature cystic teratoma (Fig. 5).

No neurological complications developed postoperatively, and the patient's symptom of urinary incontinence was improved 2 weeks later.

DISCUSSION

The spinal teratoma is an uncommon disease. The first case of this condition was described by Virchow in 1863⁶, and Gowers and Horsley described another case 25 years later¹⁰. Only a few series studies on this condition have been reported in medical literature.

Spinal teratoma accounts for only 0.1-0.5% of all spinal tumors^{5,9}. In according to Poeze et al.¹⁰, the 31 cases of 83 cases were intramedullary type, and the most of 52 cases were intradural extramedullary type. Diastematomyelia, myelomeningocele, tethered cord syndrome, and other conditions have been reported in relation to spinal teratomas^{1,8-10}. The occurrence of spinal teratomas, not associated with dysraphism is rare and is more common among infants and adolescents than among adults^{5,10}. Several authors reported that a history of trauma or surgical interventions often precedes the clinical presentation of this condition in the adult cases^{3,11}. However, our patient did not show any evidence of spinal dysraphism during physical and radiologic examination. Also, he had not previously undergone any spinal surgery or procedure.

Several authors have reported that the thoracolumbar

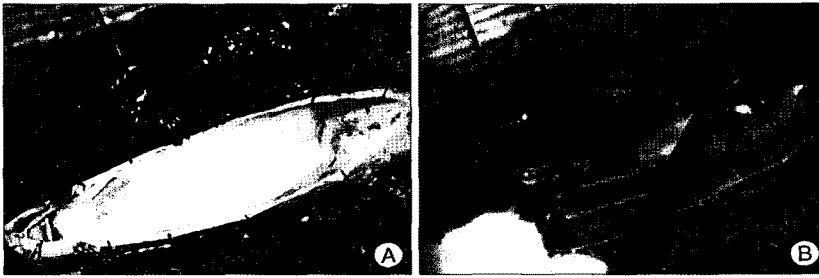


Fig. 4. Intraoperative photograph after durotomy. The yellowish fatty cyst is found and the needle-aspiration is done (A). The distal fibrous solid mass is adhered strongly to the conus medullaris and the neural filament and the gray cystic mass is noted (B).

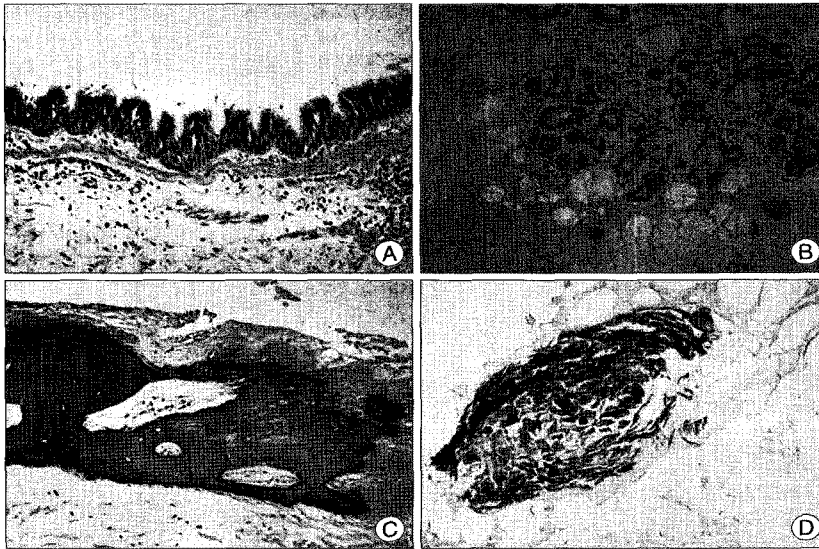


Fig. 5. Respiratory epithelial cells (endodermal origin, A), enteric glandular cells (endodermal origin, B), mature fat cells (mesodermal origin, C), bone components (mesodermal origin, C), keratinous squamous epithelial cells (ectodermal origin, D) are found.

region is most commonly affected, particularly in the area of conus medullaris^{1,8-10}. The clinical features, including weakness of the leg, sensory changes, and reflex abnormalities, are related to the location of the tumors¹⁰. In the literature, most of cases have presented with these symptoms^{1-3,6,9-11}. With regard to the clinical manifestations of the tumor in our case, the patient didn't have above-mentioned neurological abnormalities, and his chief complaint was only urinary incontinence. The operative findings revealed that the degree of spinal cord compression by the mass was severe. However, the patient's neurological symptoms were not severe, presumably because of the slow growing nature of the tumor.

Histopathologically, teratomas are composed of the remnants of all 3 germ cell layers^{1,3,5-7,9,10}. They are classified into 3 types : mature, immature and malignant teratomas. Mature teratomas mainly contain mature elements such as

cartilage, squamous epithelial cells, glands, mucosal tissue, and neural elements. On the other hand, immature teratomas are aggressive tumors, comprising primitive, undifferentiated components that resemble fetal tissues; these tumors tend to recur frequently. Malignant teratomas are derived from the yolk sac or endodermal sinus, and especially, malignant teratomas, along with the high levels of serum α -fetoprotein, are associated with a poor prognosis^{1,7}. In the classification of spinal teratomas, several authors have used numerous terms interchangeably and, in certain cases, erroneously; these terms include "epidermoid," "dermoid," "enterogenous," "teratoid," and "teratomatous" cysts⁹. However, it is essential that elements of the 3 germ cell layers are identified for it to be diagnosed as a spinal teratoma.

Several hypotheses have been proposed with regard to the origin of spinal teratomas. The most widely accepted theory is that pluripotential embryonic cells are misplaced into the dorsal midline during their normal migration from the primitive yolk sac to the gonadal ridges^{1,7,8,11}. Consequently, the primitive germ cell layers are misplaced during the early weeks of embryogenesis. This theory

has been proposed based on the fact that spinal teratomas are often located along the midline and are commonly associated with spinal dysraphism. This association suggests that a developmental fault arose during embryogenesis related to dysfunction of several factors that probably involve genetic and cellular inductive interactions^{1,7,11}.

In the diagnosis of spinal teratomas, the role of plain radiography is limited to detecting changes in the vertebral bodies, such as the erosion and widening of the interpedicular space due to the presence of a mass in the spinal canal, with or without vertebral abnormalities, including spinal bifida, vertebral body fusion or asymmetry, and diastematomyelia, at the level of the lesion⁸. A CT scan may show variable tumor density or calcification. MRI is regarded as the gold standard diagnostic technique that can reveal the location of teratomas and, consequently, the

degree of spinal cord involvement. The finding of mixed high- and low-intensity signals reflects the cystic and solid compositions of the tumor⁶.

Total surgical resection is the treatment of choice for symptomatic patients. However, intimal adhesion of the teratomas to the surrounding neural parenchyma is observed about 50% cases^{1,2,8,9}, so it is difficult to resect tumors totally. Several authors reported that subtotal resection of these tumors increases the chance of recurrence, so they recommend that it is important to resect the tumor as much as possible, while preserving all the surrounding neural tissue^{8,9,11}. In addition, it is important not to spill the cystic contents into the intradural space during surgery, because it can also lead to the development of aseptic chemical meningitis with or without obstructive hydrocephalus^{3,7,11}. The role of adjuvant therapies, including radiotherapy and chemotherapy for the remnant tumors, has not been characterized clearly⁸. Its effects for immature and malignant teratomas also remain controversial^{2,5}.

CONCLUSION

We report a rare case of a spinal intradural extramedullary mature cystic teratoma in an adult. In our case, we performed a subtotal removal, due to severe adhesion of the teratomas to the surrounding normal tissue. Therefore, we believe that it is necessary to follow up the consecutive radiologic images, and the symptoms and neurological changes should be observed strictly during long term follow-up.

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