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Case Report

Adult onset tethered cord syndrome associated with intradural dermoid cyst. A case report

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Study design: A case report and a review of literature.
Objectives: To describe a rare case of adult onset tethered cord syndrome associated with intradural dermoid cyst.
Setting: General Orthopedics, Japan.
Methods: A 50-year-old woman was referred to us because of right leg pain and pollakiuria. Neurological examinations and radiological assessments including myelography, computerized tomography scan and Magnetic resonance image were carried out. We diagnosed it as the adult onset tethered cord syndrome associated with an intradural cystic lesion.
Results: The cystic lesion was totally removed following laminectomy from L5 through S4. Histologically, the tumor was diagnosed as a dermoid cyst.
Conclusions: Intradural dermoid could produce adult onset tethered cord syndrome, but it was not reported in the English literatures to our knowledge.
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Keywords: intradural dermoid cyst; adult onset; tethered cord syndrome

Introduction

The symptoms of tethered cord syndrome (TCS) cases mostly appear during infancy and childhood,[1–3] but rarely noticed for the first time after the third decade.[4,5] Intradural dermoid cysts have been rarely reported among intraspinal tumors.[6–9] In this report, we describe a rare case of adult onset TCS associated with an intradural dermoid cyst.

Case report

A 50-year-old woman visited our hospital with a 6-year history of low-back pain, right leg pain and pollakiuria. The right leg pain seemed to be along the right sciatic nerve and increased gradually. Physical examination showed localized tenderness, dimpling and tufts of hair in the sacral region. Except for decreased sensation to pinprick over her right leg, neurological examinations were normal including the anal tonus, the deep tendon reflexes and muscle strength.

Plain radiographs showed the defect of the posterior elements of the sacrum and scalloping of the sacral bodies. A myelogram suggested an intradural mass from the fifth lumbar body to second sacral body level, and computerized tomography (CT) after myelography showed a thickened filum at L5 (Figure 1). Magnetic resonance image (MRI) showed that the mass was low-signal intensity on the T1- and high-signal intensity on the T2- weighted image heterogeneously. The rim of the mass was only enhanced by gadolinium (Figure 2). The conus medullaris was found at L5, indicating the tethering by the thickened filum terminale. The angiography did not provide any new information. These findings suggested that the symptoms were derived from adult onset tethered cord syndrome associated with spinal cystic condition.

The patient underwent surgery to remove the spinal cyst with dermal sinus in December 1999. There was direct connection between the skin sinus and the intradural cyst. Following laminectomy from L5 to S4 and opening of the dura mater, a cystic yellow mass was found among the cauda equina, and all sacral roots were compressed rightward. The tip end of the conus was unclear, but the cyst was not connected with the neural tissues. The intradural cyst was completely removed, and contained granules of white cholesterol. The conus medullaris, cauda equina and filum terminale became loose after the removal; thus, the thickened film

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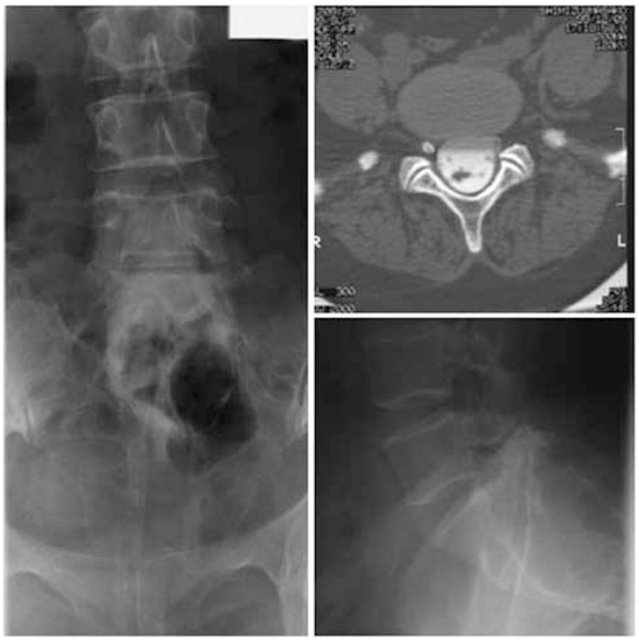

Figure 1 Left: Myelography showing dural ectasia and contrast pooling which obliterates intradural details. Right/upper: CT scans after myelography at L5 clearly showing the abnormally low conus dorsal to the lumbosacral nerve roots. Right/bottom: Plain radiographs showed the defect of the posterior elements of the sacrum and scalloping of the sacral bodies

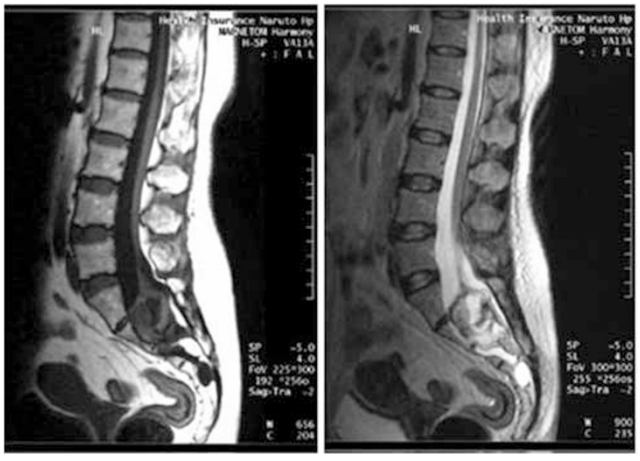

Figure 2 MR image of the lumbosacral regions demonstrated a heterogeneous intradural lesion extending from S1 to S3 level. The mass of low-signal intensity on the T1 weighted and high-signal intensity on the T2 weighted. The conus terminated intradural mass at L5

terminale was not resected. The dural defects were closed with lumbosacral fascial grafts.

In the histological examination (Figure 3), the lining of the cyst consisted of stratified squamous epithelium with hair follicles, sebaceous and sweat glands, and the fat organization was at a much deeper area. These findings were consistent with a dermoid cyst.

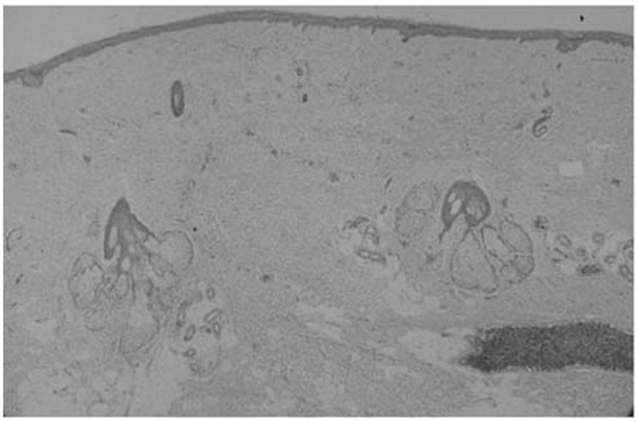

Figure 3 Histopathologic specimen of a cyst lined with squamous epithelium with hair follicles, sebaceous and sweat glands. HE ($\times 50$)

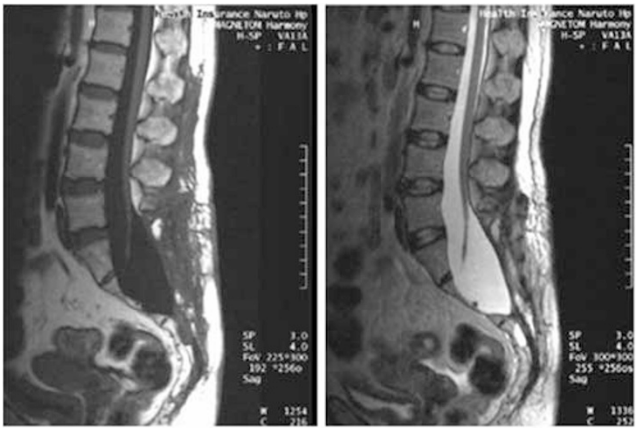

Figure 4 Postoperative image demonstrating that low-placed conus remains, although the dermoid cyst was extracted completely

At one year after the surgery, the patient reports mild posterior thigh ache, minor difficulty in urination and slight perineal numbness. An MRI after the surgery demonstrated the conus at the same level in the spinal canal and no residual cyst (Figure 4).

Discussion

We described here an intradural dermoid cyst showing adult onset TCS. The lining of dermoid cysts includes skin appendages such as hair follicles and sebaceous or sweat glands in addition to stratified squamous epithelium. A diagnosis of this type of tumor is easy when histopathologically studied. Dermoid cysts have been reported to be very rare in the central nervous system. Goodrich *et al*[6] reported only 10 dermoid cysts out of 979 intraspinal tumors, accounting for just over 1%. Other reports also indicated the incidence to be about 0.5–2%.[7–9] In the spinal canal, dermoid cysts commonly appear in the lumbosacral spinal canal, which was the

site of the present case. Concerning age, this type of tumor is frequently encountered at younger ages than epidermoid cysts according to the review by Guidetti and Gagliardi.[7] Shikata *et al*[10] reported that the mean age of patients at the time of surgery was 1.5 years old for dermoid cysts and 36.5 years old for epidermoid cysts. Those previous reports indicated that dermoid tumors are usually found in young people. However, in the present case, the tumor was first noticed at the age of 50, which is quite rare.

As for the pathogenesis of the dermoid cysts, two theories have been proposed; that is, abnormal fetal development,[11] and trauma inflicted after birth.[12] According to the former theory, a migrant ectoderm becomes enclosed within the neural tube during the third to fifth week of embryonic life, causing epidermoid or dermoid cysts, or other related malformations such as a congenital dermoid sinus. The latter theory suggests that cysts develop because skin tissue is forced into the vertebral canal during lumbar puncture, trauma or surgery. This patient, who had never undergone lumbar puncture or injury, and who had a congenital anomaly of low-placed conus, appears to have had abnormal fetal development. This occurs very rarely in adults.

Patients with tethered cord syndrome rarely have symptomatic onset in adulthood. Adult onset TCS presents very different symptom complexes[9,13] compared to childhood onset TCS. Pain is uncommon in children, but is common in adults. The character of the pain is also different in the two age groups. In children, it is located in the lumbosacral regions with only variable radiation to the legs. In adults, the pain is dysesthetic, diffuse and most common in the legs and perineal region. Progressive cavovarus deformity of the foot is one of the most common features in children with tethered cord syndrome. None of the adult patients who did not have pre-existing foot deformity developed this problem with adult onset of neurological dysfunction. The incidence of urological symptoms appears to be similar in the childhood and adult syndromes. Growth causes the onset of symptoms in children. In contrast, various factors, including injury, mechanical stress, spondylosis, disc herniation and spinal canal stenosis, cause these symptoms in adults. These factors had nothing to do with the onset of this case. Pathological entities causing the tethering are various. Adults usually have a higher ratio of lipoma and lipomeningocele than children.[1] Another report ascribed intrasacral meningocele as the cause.[14–16]

Dermoid cysts are essentially benign, but will recur if incompletely removed. They will become enlarged if diagnosis and medical treatment are not performed. The rupture of dermoid cysts and the migration of free-fat globules may cause a series of complications. Among the complications, the literature reports a case of obstructive hydrocephalus and aseptic meningitis.[17,18] Pang and Wilberger[13] report bladder dysfunction in adult tethered cord syndrome patients that are equally disappointing, but spastic small-capacity bladders tend to have a better recovery potential than the atonic bladder. Early and accurate diagnosis and complete resection under microscope are of prime importance.

In conclusion, in this case report, we described a very rare case showing adult onset tethered cord syndrome associated with intradural dermoid cyst. The MRI was very useful for early and accurate diagnosis. The presence of this kind of tumor must be taken into account in cases with adult onset TCS.

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