CASE REPORT

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Congenital Elongated Lumbar Dermoid Cyst Combined with Sacral Meningocele

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Dermoid cysts of the spinal canal are rare benign congenital tumors, accounting for 1~2% of all intraspinal tumors. We report a case of lumbar extramedullary cyst, combined with congenital sacral meningocele. The clinical features, characteristics on MRI, pathologic findings, and surgical treatment of such a rare extramedullary benign tumor is discussed with the relevant literature.

KEY WORDS : Dermoid cyst · Lumbar spine · Meningocele.

Introduction

Dermoid cysts are rare, and account for 0.7~1.8% of tumors in the central nervous system[1]. Spinal dermoid cysts are even rarer, constituting 1~2% of intraspinal tumors. Spinal dermoid cysts can be intramedullary, intradural extramedullary or extradural (25% intradural : 75% extradural)[16]. They occur predominantly in the lumbosacral region (60%), involving the cauda equina and conus medullaris, and are relatively rare in the upper thoracic (10%) and cervical regions (5%)[7]. Like epidermoid cysts, dermoid cysts have a lining of stratified squamous epithelial cells; however, they also contain dermal structures, such as sebaceous and/or sweat glands, hair follicles, and even teeth[2], and calcifications commonly occur. Intraspinal dermoid cysts are frequently associated with mesodermal malformations. This is particularly true when the lesion involves the vertebrae (for example hemivertebrae, absent vertebrae, fused vertebrae, butterfly vertebrae, or midline bony spurs), and when the lesion is associated with dermal sinuses, myelomeningocele, syringomyelia, and repeated lumbar punctures[3,12,14]. Compared with true neoplasms, which grow by progressive cell division, intraspinal dermoid cysts enlarge by desquamation of normal cells and secretion of dermal elements into a cystic cavity well circumscribed by a thick wall of connective tissue. The clinical history is related to the lesion site and/or mass effect. Because of their slow rate of growth, dermoid cysts can reach a considerable size without causing any symptoms. As the cyst increases in volume, it conforms in shape to any available subarachnoid space and tends to become tightly adherent to neighboring structures[11,17]. MRI has recently become the method of choice for examination of the spinal cord and early diagnosis of these tumors is now possible. Here we report the rare occurrence of congenital elongated lumbar dermoid cyst combined with sacral meningocele.

Case Report

A 48-year-old female had suffered from low back pain for 10 years, and had hypesthetic pain in the left leg, saddle numbness, and difficulty in voiding for one year prior to presentation. T1- and T2-weighted MRI of the lumbar spine revealed a lumbar spinal tumor. She was admitted to the neurosurgical department for surgical decompression.

Her past history was neither specific to medicine nor to acupuncture procedure. She was aware of meningocele after birth, but did not pursue a follow up study because no specific neurologic symptoms. Physical examination revealed a 4×3cm sized mass in the central to right sacral skin area, hypesthetic pains in the left L4/5 dermatomes, and difficulty in voiding, but no motor weakness. Neurological intermittent claudication

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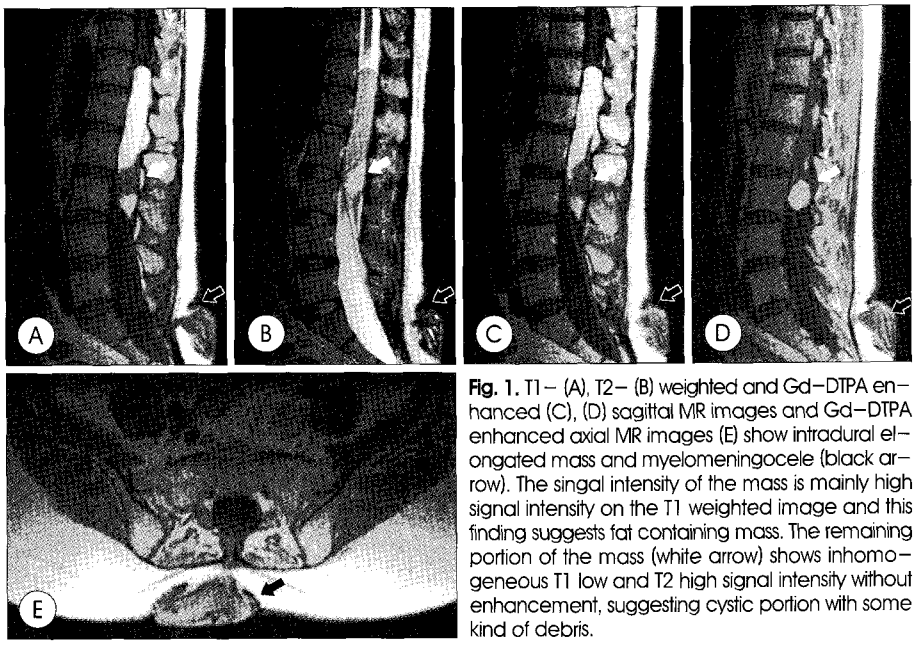

Fig. 1. T1- (A), T2- (B) weighted and Gd-DTPA enhanced (C), (D) sagittal MR images and Gd-DTPA enhanced axial MR images (E) show intradural elongated mass and myelomeningocele (black arrow). The singal intensity of the mass is mainly high signal intensity on the T1 weighted image and this finding suggests fat containing mass. The remaining portion of the mass (white arrow) shows inhomogeneous T1 low and T2 high signal intensity without enhancement, suggesting cystic portion with some kind of debris.

amounted to fifty meters, and straight leg rising test showed a 45° limit in the left leg. Sphincter function was objectively normal.

MRI demonstrated multiple, well-circumscribed, elongated intradural extramedullary masses extending to the compressed thecal sac and shifting the conus medullaris from L1 to T12 level. MRI studies of dermoid tumors give a varied appearance depending on the fat contents of the tumor. T1-weighted MRI showed the mass to be iso-signal to low-signal intense in the upper portion, but high-signal intense in the lower portion. T2-weighted MRI showed the mass to be slightly high-signal intense in upper portion, but low-signal intense in the lower portion. T1-weighted MRI with a gagolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA)-enhancing study showed no enhancement, but revealed a congenital meningocele in the sacrum. The trace of the tract was seen between thecal sac and meningocele, but was not connected. This is presumed at a regression change of the tract (Fig. 1).

Based on these observations, a total laminectomy was performed on the L2 level with the upper portion of L3. The dura was exposed and incised vertically, exposing a well capsulated whitish mass. The capsule was dissected free from the neural tissue, and an opening in the capsule was made to allow partial removal of the soft creamy contents. After partial removal of the capsule, additional lipid droplets were found; these were irrigated and aspirated. The pathologic report on the frozen sample and the final report confirmed the diagnosis of dermoid cyst. And skin mass was pathologic confirmed the diagnosis of meningocele.

The post-operative course was uneventful and without neurological deterioration. MRI performed on post-operative day 7 confirmed decompressed neural elements. Voiding difficulty, saddle numbness and left leg pain were improved at discharge.

Discussion

Dermoid cysts account for 0.7 ~1.8% of tumors in the central nervous system with a cranial to spinal ratio of 6 : 1[1,12]. Dermoid cysts constitute 13~17% of all primary spinal cord tumors between 1 and 15 years of age[4,10], and generally present at an average age of 15 years with a female predominance[10]. According to Dias and Walker, the pathogenesis of spinal epidermoid cysts and dermoid cysts is mainly congenital, originating from the epithelial tissue displaced during the closure of the neural tube between the 3rd and 5th weeks of gestation[5,6]. Diagnosis of dermoid and epidermoid cysts is usually based on the characteristic nature of their contents revealed by histological examination. Dermoid and epidermoid cysts are lined with stratified squamous epithelium supported by an outer layer of collagenous tissue. Progressive desquamation of keratin from the epithelial lining toward the interior of the cyst produces a soft white material[8]. Differentiation between the two forms is based on the presence of skin adnexa in dermoid cysts only. Although calcification is rare in epidermoid tumors, approximately 20% of dermoid tumors show evidence of calcium deposition[9]. In the present case, histologic findings showed a keratinized squamous epithelial lining, and sebaceous glands including hair follicles. High power microscopy showed the contents of the cyst to comprise keratinous material and a few hairs. The cyst was lined with squamous epithelium and the wall showed dystrophic calcification (Fig. 2).

Although they grow more quickly than epidermoid cysts, the slow growth of dermoid cysts often leads to a delay in their diagnosis and symptoms may persist for many years before diagnosis. Almost 50% of patients with these tumors have associated congenital anomalies[13]. These lesions may also occur after the introduction of dermal elements into the spinal subarachnoid space due to lumbar puncture, surgery or trauma. Our patient had no history of a lumbar puncture, no definite congenital bone anomalies and no definite connected spinal dysraphism, but did have an independent meningocele in the sacral region. Therefore, it was presumed that the dermoid cyst of this patient was congenital in origin. Although the signal

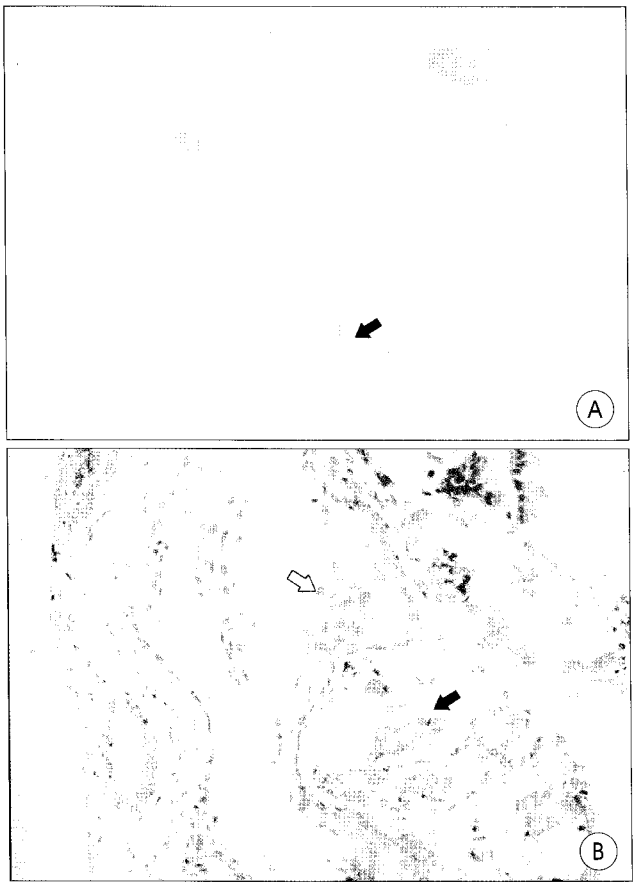

Fig. 2. A : Cyst contents consisted of keratinous material and a few hairs (black arrow) (H&E, X400). B : The cyst is lined by squamous epithelium (white arrow) and the wall shows dystrophic calcification (black arrow).

intensity of dermoid cysts varies, it is typically iso-signal or slightly high-signal intense compared with that of CSF on all sequences and shows no contrast enhancement. This variability in signal characteristics, which might be related to the chemical state of cholesterol or the relative composition of cholesterol and keratin, makes pre-operative diagnosis difficult. In our case, T1-weighted MRI showed the mass to be iso-signal to low-signal intense in the upper portion, but high-signal intense in the lower portion. T2-weighted imaging showed it to be slightly high-signal intense in the upper portion, but low-signal intense in the lower portion. T1-weighted MRI with a Gd-DTPA enhanced study showed no enhancement. The inhomogeneous signal on long relaxation time/long echo time images from the more solid core of the mass reflected a mixture of intact and partially degenerated materials of hair, glandular elements and more solid cholesterol[15]. However, other cystic spinal tumors with fluid content could not be ruled out (Fig. 1).

Conclusion

Spinal dermoid cysts are rare. There are typically no symptoms because of the slow rate of growth and no compressed neural elements in the early stage. In the case described here, the patient had some neurologic symptoms, which necessitated further study and proper management. We report the rare occurrence of a congenital lumbar dermoid cyst combined with sacral meningocele that was successfully managed by surgical removal of the cyst.

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