Intradural Extramedullary Epidermoid Cyst at the Conus Medullaris level with Thoracic Syringomyelia: a Case Report

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ABSTRACT

Spinal epidermoid cysts are benign tumors. Syringomyelia secondary to intramedullary tumors are frequently observed. However, the association between syringomyelia and spinal intradural extramedullary epidermoid cyst in the conus medullaris region is extremely rare. We present the case of a 3-year-old male who was admitted with paraparesis and urinary retention. Magnetic resonance imaging (MRI) of the spine demonstrated intradural extramedullary lesion, compatible with epidermoid cyst, that at the conus medullaris level and a large syringomyelia extending from T4 to L1 vertebrae. Total microsurgical excision of the cyst was performed. No additional drainage was carried out for the syringomyelic cavity. Histopathological examination verified the diagnosis of the epidermoid cyst. Total excision of the cyst and disappearance of the syringomyelia were observed on MRI at 15 days postoperatively. We have clarified the etiology, clinical, histopathological and radiological features, differential diagnosis, and treatment modalities of spinal epidermoid cysts. In addition, we have discussed the possible mechanisms of syringomyelia formation in spinal intradural lesions.

KEYWORDS

conus medullaris; epidermoid cyst; syringomyelia; spine

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INTRODUCTION

The term "inclusion cysts" includes dermoid and epidermoid tumors/cysts. They are derived from cutaneous ectoderm (1). Spinal epidermoid cysts are rare and slow-growing benign lesions. They constitute <1% of all spinal tumors (2, 3). Most of the spinal epidermoid cysts are located in the lumbosacral and thoracic regions (4). Conus medullaris epidermoid cysts are extremely rare entities.

Syringomyelia is characterized by the presence of cystic cavities inside the spinal cord. It is a secondary process with many etiologies. Chiari malformations, neoplasms, arteriovenous malformations, arachnoiditis, and spinal dysraphisms are some examples (5). Syringomyelia associated with intramedullary tumors are frequently observed. The most common tumors associated with syringomyelia are astrocytomas and ependymomas. In addition, their most common locations are the lower cervical and upper thoracic regions (6, 7). However, extramedullary tumors at the conus medullaris level associated with syringomyelia are uncommon coexistences. Here we describe and discuss the case of a patient with conus medullaris epidermoid cyst associated with a large thoracic syringomyelia.

CASE REPORT

A 3-year-old male was admitted with increasingly leg weakness that had persisted for 2 months. Neurologic examination revealed that the proximal and distal muscular forces in his right and left lower extremities were 3/5 and 2/5, respectively. A large syringomyelia, extending from T4 to L1 vertebrae, was detected on thoracic magnetic resonance imaging (MRI). In lumbar MRI, T1-weighted imaging demonstrated isointense expansion at the L1-2 vertebrae level. T2-weighted imaging revealed an intradural extramedullary regular lesion that had hyperintense capsule-like formation with isointense content and that excessively compressed the conus medullaris. It was approximately 15 × 15 mm in size. T1-gadolinium-enhanced imaging showed that dense cystic mass had circumferential contrast enhancement. Besides, some cauda equina fibers had contrast enhancement (Figure 1). In addition, renal ultrasonography was performed before and after micturition. Significant post-micturition residue was determined. He underwent surgery. Total microsurgical excision of the lesion was achieved with the help of neuromonitoring after L1 and L2 laminotomies. Also, the integrity of the facet joints was protected. After dural opening, the lower end of the spinal cord (conus medullaris) and the beginning of the cauda equina fibers appeared compressed by an anterior extramedullary lesion. Dense pearly content and whitish cyst capsule were completely excised. No additional drainage procedure for the syringomyelic cavity was considered at this surgical intervention. Besides, CSF leakage was not observed from the neighbouring syringomyelic cavity after cyst removal. In the closure, L1 and L2 laminoplasties were performed. In the early postoperative period, muscular force in his right and left lower extremities had improved to 4/5 and 3/5, respectively. Histological examination of the specimens demonstrated that the cyst walls were lined with stratified squamous epithelium without skin adnexa. Furthermore, desquamation of keratin from the epithelial lining was detected. This examination revealed epidermoid cyst (Figure 2). There was no finding of residual tumor on MRI at 15 days postoperatively. In addition, disappearance of the syringomyelia was seen. However hyperintensity at T2 weighted image compatible with myelomalacia, existed after the cyst resection, was observed (Figure 3A,B,C). At 20 days postoperatively, renal ultrasonography was performed before and after micturition, which showed decreased post-micturition residue. Physical therapy and rehabilitation were indicated at 1 month postoperatively. In the second month control his examination revealed no neurological deficits. Besides, significant decrease of myelomalacia was seen at the second month follow-up MRI. (Figure 3D).

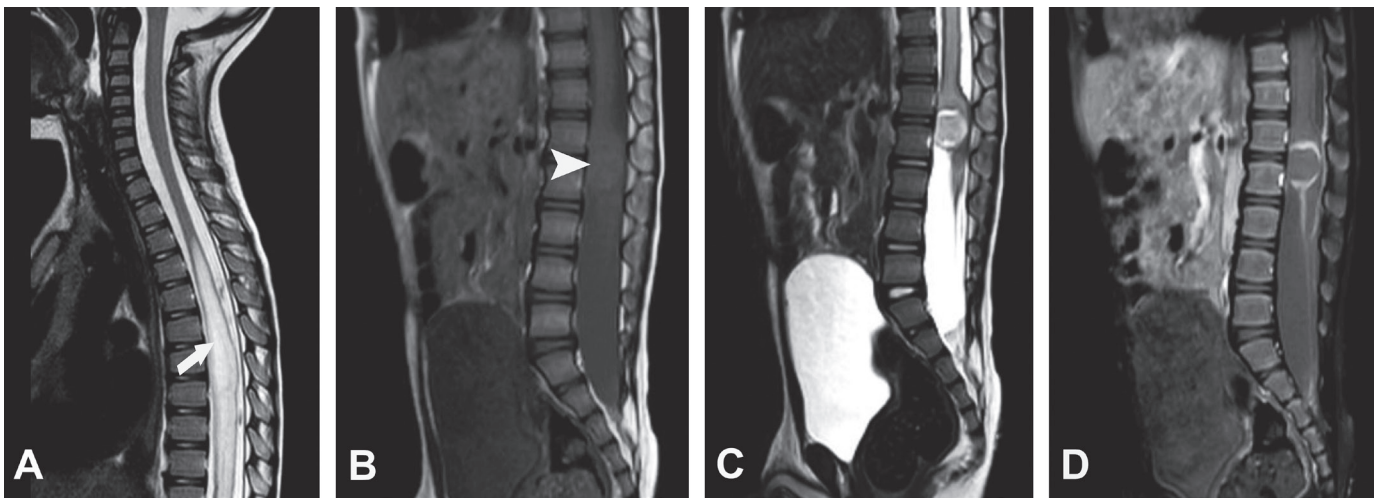

Fig. 1 In the preoperative (A) thoracic MRI, a syringohydromyelia (arrow), extending from T4 to L1 vertebrae, was observed. Lumbar MRI (B) T1-weighted sagittal image showed hypointense expansion (arrowhead) at the L1-2 vertebrae level. (C) T2-weighted image showed a lesion that had hyperintense capsular formation with isointense content and that excessively compressed the conus medullaris. (D) T1-gadolinum-enhanced image demonstrated that dense cystic mass had circumferential contrast enhancement. Besides, some cauda equina fibers had contrast enhancement.

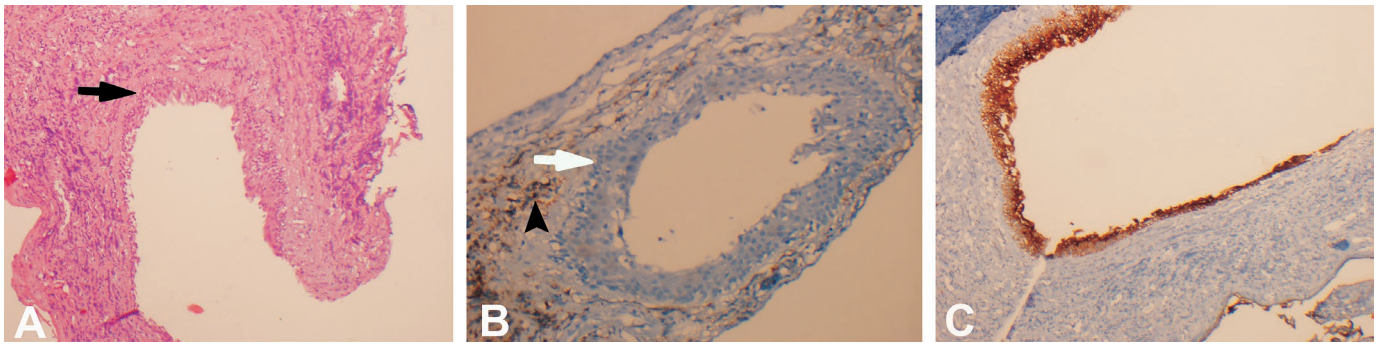

Fig. 2 In the histopathological examination, (A) cystic lesion with stratified squamous epithelium (black arrow) (hematoxylin–eosin, ×100). (B) Inflammatory cells (arrowhead), stained by leukocyte common antigen (LCA), around the cystic lesion with stratified squamous epithelium (white arrow) (LCA, ×100). (C) Stratified squamous epithelium stained with pancytokeratin. Desquamation of keratin from the epithelial lining can be observed (pancytokeratin, ×100).

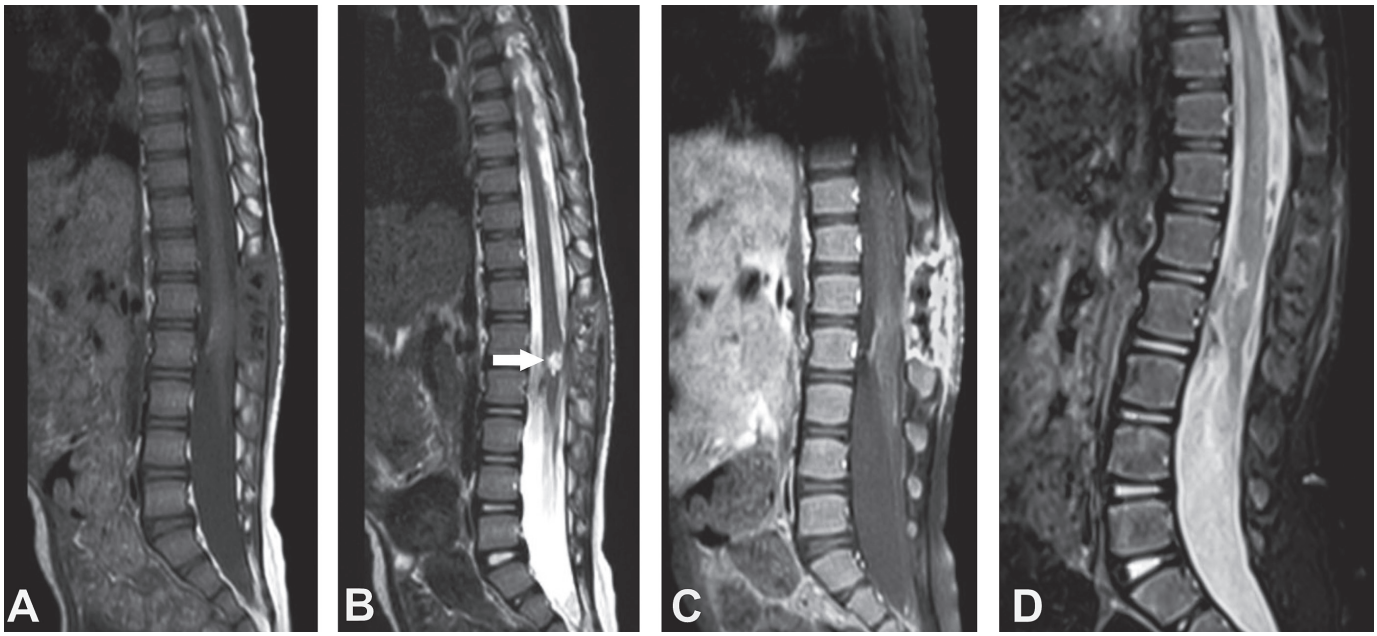

Fig. 3 Postoperative MRIs. Disappearance of the syringohydromyelia was observed. There was no finding of residual tumor. Decompression of the conus medullaris and cauda equina fibers is shown. (A) T1-weighted, (B) T2-weighted (arrow: hyperintense image thought to be myelomalacia due to chronic compression, which existed after the cyst resection), and (C) contrasted sagittal section at 15 days postoperatively. (D) T2-weighted sagittal section also showed significant decrease of myelomalacic aspect at 60 days postoperatively.

DISCUSSION

Spinal epidermoid cysts may be acquired or congenital. Acquired epidermoid cysts, due to iatrogenic penetration of skin fragments, have been frequently reported after lumbar puncture or meningomyelocele repair with years of latency (1, 8, 9). Congenital spinal epidermoid cyst is thought to be an inclusion of ectodermal tissues during closure of the neural tube between the third and fourth weeks of embryonic development (3). Occult spinal dysraphisms, spina bifida aperta, hemivertebra, and dermoid tracts are often associated with congenital cysts (1, 3, 8). Our patient had no known history of meningomyelocele repair such as open wound trauma or lumbar puncture; therefore, the tumor was thought to be congenital.

Signs and symptoms arising from epidermoid cysts vary with the extent of involvement and are not different from other spinal space-occupying lesions. Paraparesis, sensory loss, urological manifestations, pathologic reflex appearance, deep tendon reflex changes, and back/extremity pain are usually found at presentation. The slow growth of epidermoid cysts often leads to a delay in their diagnosis (2).

MRI is useful for diagnosis, particularly contrast-enhanced studies, which also show the exact location and extension of the lesion. MRI demonstrates that epidermoid cyst is hypointense on T1-weighted and hyperintense on T2-weighted images (8). Peripheral postgadolinium enhancement with well-defined limits without perilesional edema contributes to diagnosis. Some authors emphasize that peripheral enhancement represents normal peripheral surrounding tissue reaction, whereas others consider it as an outer capsule composed of tumor cells (9). In this patient, circumferential contrast enhancement was due to capsular formation. However, contrast enhancement of the some cauda equina fibers might be reactional. During

the operation, we observed conspicuous capsular formation; therefore, we obtained tissue samples as well as the capsule-like formation and dense content of the cyst for histopathological examination.

Ependymomas are the most common space-occupying lesions at the conus medullaris–cauda equina level. Lymphomas, astrocytomas, gangliogliomas, and ganglioneuromas can be considered during differential diagnosis (7, 10, 11). They can be frequently identified and distinguished by MRI studies. Astrocytomas, gangliogliomas, and ganglioneuromas have heterogeneous contrast patterns. Ependymomas have homogenous contrast enhancement, and epidermoid cysts have peripheral contrast enhancement (7, 9, 11). Nevertheless, radiological differentiation is sometimes difficult for these spinal lesions, but they can be histopathologically diagnosed.

Histopathological examination is critical for diagnosing of epidermoid cysts. Differentiation between the two forms of inclusion tumors (dermoid/epidermoid) is based on the presence of adnexal skin structures, such as sebaceous glands, eccrine glands, and hair follicles. They are present only in dermoid cysts (1, 2). A specific histological feature of the epidermoid cyst is a lined stratified squamous keratinizing epithelium surrounded by an outer layer of collagenous tissue and/or inflammatory cells without skin adnexa. Desquamation of keratin from the epithelial lining generates cholesterol crystals (2, 9).

Total excision including the capsule is the curative treatment for spinal epidermoid cyst because it is a benign condition. Sometimes, because of its anatomical features, intramedullary tumor shows tight adherence to the arachnoid membrane and spinal cord, thereby causing some surgical difficulties for safe and complete resection. Therefore, subtotal resection is also commonly performed to avoid neurological deficits (1, 2). Local recurrence is reported especially after subtotal excision. Incomplete excision of basal germinal cells of the tumor is suggested to be the cause of recurrence (12). Long-term follow-up is necessary for tumor recurrence risk. Surgeons should keep in mind that cyst content includes fat and cholesterol. These can cause an inflammatory reaction that leads to aseptic chemical meningitis (Mollaret's meningitis) (8). Plugging of proximal and distal arachnoid spaces and irrigating the site with normal saline prior to dural closure reduces the risk of chemical meningitis (9). We performed total excision in this case. Besides, the patient did not develop any inflammatory reactions and neurological deterioration postoperatively.

The exact pathogenesis of syringomyelia accompanied with spinal cord tumors is unclear. Different theories have been proposed. Some of them are intramedullary softening due to an impaired blood circulation, stasis of tissue fluid resulting from occlusion of the drainage pathways, effect of edema, and spontaneous autolytic reactions of the tumor (7, 13). Another suggested mechanism of syrinx formation was obstruction of cerebrospinal fluid (CSF) flow (13, 10, 14). Given these possible reasons, syringomyelia associated with intramedullary tumors can be frequently observed. However, extramedullary tumors associated with syringomyelia are very rare. In our case, syringomyelia most likely developed secondary to the mass effect of the epidermoid cyst. It has been postulated that cyst may cause blockage of CSF flow dynamics. Accordingly, excision of the cyst can remove the mass effect and regenerate these CSF flow alterations. Hence, after the total resection of the cyst, disappearance/reduction of the syrinx cavity was achieved, in spite of CSF leakage was not observed from the neighbouring syringomyelic cavity. Besides, no additional drainage procedure for the syringomyelic cavity was considered.

In conclusion, spinal intradural extramedullary epidermoid cyst at the conus medullaris level associated with thoracic syringomyelia is a very rare entity. MRI is very useful for diagnosis, identifying the exact location of the cyst, and detecting comorbidities. The clinical course is dependent on the neurological condition of the patient at presentation and any delay in the diagnosis. Moreover, appropriate early surgical intervention with total excision results in good prognosis. Syringomyelia most likely developed secondary to the mass effect of the cyst and blockage of the CSF flow dynamics. Therefore, excision of the cyst can ensure the disappearance or reduction of the syringomyelia.

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