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Thoracic Syringomyelia with Intradural Extramedullary Epidermoid Cyst at the Conus Medullaris level: a case report

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Abstract: The work aims to present a case from practice where an extramedullary tumor of the cauda equina roots caused the formation of a large syringomyelic cyst of the thoracic spinal cord, which is confirmed by magnetic resonance imaging (MRI). Moreover, neurological disorders in the patient prevailed from the thoracic spinal cord. After an operation to remove a tumor (epidermoid cyst), thoracic syringomyelia resolved, and neurological disorders regressed with it, which is confirmed by electroneuromyography of the lower extremities. The MRI picture also marks a complete regression of the syringomyelitics cavity.

Keywords: cauda equina root tumor, syringomyelia, an epidermoid cyst.

Relevance

The term "inclusion cysts" refers to dermoid and epidermoid cystic tumors. They are formed from the skin ectoderm (1). Epidermoid cysts of the spine are rare and slow-growing benign tumors. They account for less than 1% of all spinal tumors (2, 3). Most epidermoid cysts of the spine are located in the lumbosacral and thoracic regions (4). Epidermoid cysts of the conus of the spinal cord are extremely rare. Syringomyelia is characterized by the presence of cystic cavities within the spinal cord. This is a secondary process that develops for various reasons. Malformations of the craniovertebral junction - Chiari, neoplasms, arteriovenous malformations, arachnoiditis, and spinal dysraphisms are some of them (5). Syringomyelia associated with intramedullary tumors is often observed. The most common tumors associated with syringomyelia are astrocytomas and ependymomas. In addition, their most common sites of localization are the lower cervical and upper thoracic regions (6, 7). However, extramedullary tumors at the level of the *conus medullaris* associated with syringomyelia are not typical. This article describes and discusses the case of a patient with an epidermoid cyst of the conus of the spinal cord associated with large thoracic syringomyelia.

A case presentation.

A 3-year-old boy presented with paresis in both legs, which developed within 2 months. Neurological examination showed that in the proximal and distal muscle groups, the strength in his right and left lower limbs was 3/5 and 2/5, respectively. A large syringomyelic cyst extending from T4 to L1 vertebrae was found on magnetic resonance imaging (MRI) of the thoracic spinal cord. Lumbar MRI in T1-weighted mode revealed isointense expansion at the level of the L1-2 vertebrae. T2-weighted imaging revealed an intradural extramedullary neoplasm that had a hyperintense capsule-like shape with isointense content. This tumor compressed the cone of the

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spinal cord and was approximately 15 × 15 mm in size. Gadolinium T1-enhanced imaging showed that the dense cystic mass had circumferential contrast. In addition, some cauda equina roots also had contrast enhancement (figure 1).

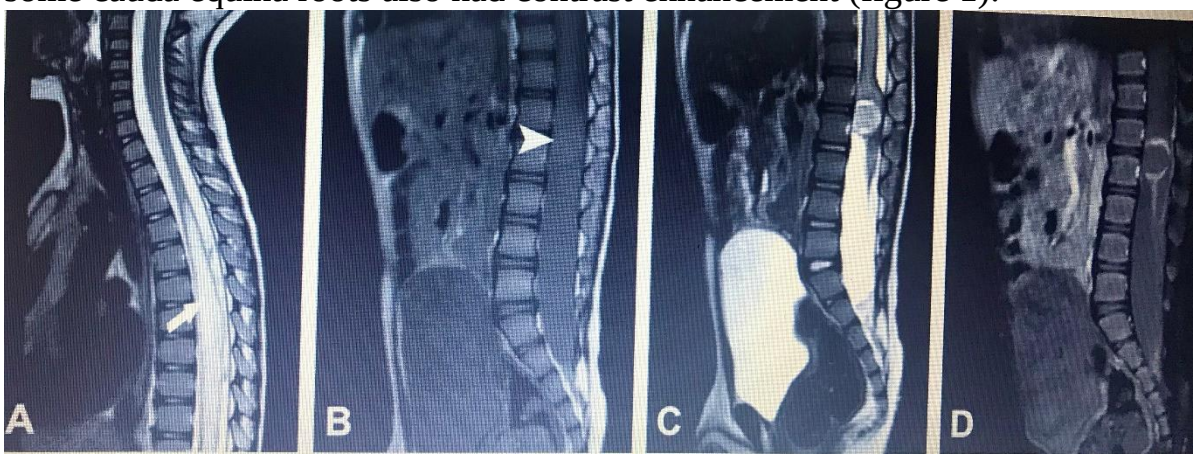

Figure 1. Preoperative (A) MRI of the thoracic spinal cord showed syringohydromyelia (arrow) extending from T4 to L1. MRI of the lumbar (B) - T1-weighted sagittal image showed hypointense expansion (arrow) at the level of the L1-2 vertebrae. (C) - T2-weighted image showed changes having hyperintense capsule formation with isointense contents and obvious compression of the spinal cord conus. In addition, some fibers of the cauda equina have contrast enhancement.

An electroneuromyographic study (ENMG) showed a decrease in the speed of impulse conduction along the neural radicular fibers segmentally along L1-2 from 2 sides with some asymmetry, lower left. Along with this, there was a moderate decrease in the speed of conduction along the efferent fibers at the level of the thoracic region Th10-12. The amplitude of the maximum M-response was reduced in the femoral, tibial, and peroneal muscle groups. Thresholds of nerve excitation were moderately increased symmetrically on both sides.

In addition, when examining the kidneys on ultrasound before and after urination, significant residual urine is determined. The patient underwent surgery. Total microsurgical removal of the cystic tumor was performed after L1 and L2 laminectomy using neuromonitoring. After opening the dura mater, it was found that the *conus medullaris* and the beginning of the fibers of the *cauda equina* were compressed by an extramedullary formation in front. Dense mother-of-pearl contents and a whitish cyst of the capsule were completely resected. No additional procedures for drainage of the syringomyelia cavity were performed during this surgical intervention. In the early postoperative period, muscle strength in the right and left lower extremities improved to 4/5 and 3/5 points, respectively. Histological examination of the tumor tissues showed that the cyst walls were lined with stratified squamous epithelium without skin appendages. In addition, desquamation of keratin from the epithelial membrane was found. The histological diagnosis was an epidermoid cyst.

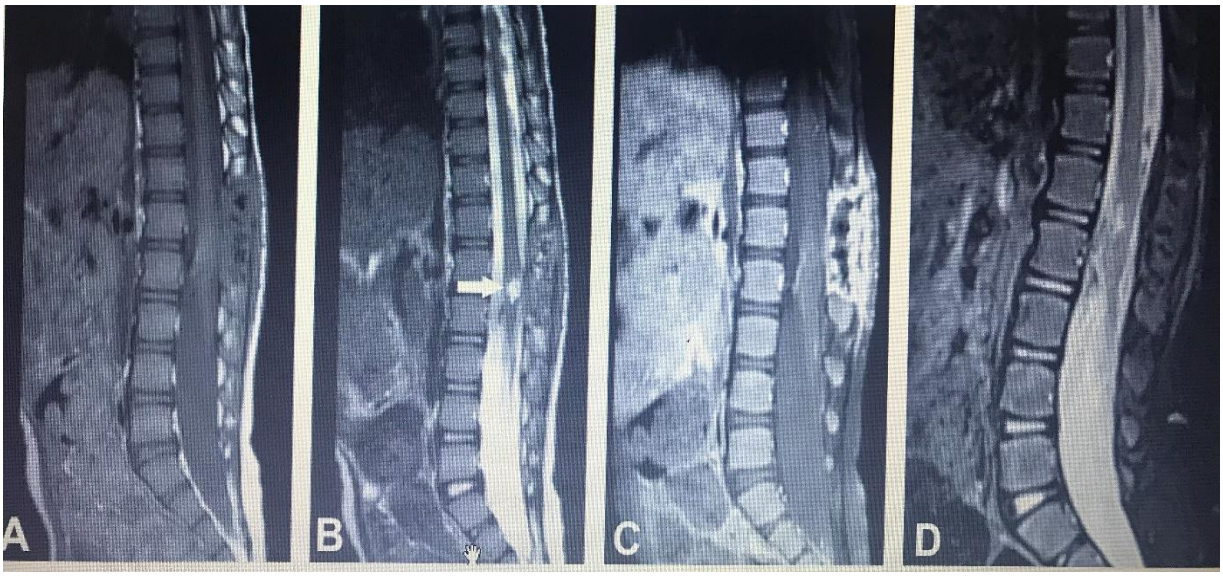

Figure 2. Postoperative MRI. The disappearance of syringohydromyelia is noted. There is no residual tumor. The medullary cone and fibers of the cauda equina are decompressed. A - T1-weighted; B, T2-weighted (arrow indicates hyperintense image, which is regarded as myelomalacia due to chronic compression that persisted after cyst resection); C - contrast sagittal section 2 weeks after surgery; D - T2-weighted sagittal section also shows a significant reduction in myelomalacia 2 months after surgery.

MRI no-showed tumor after 15 days of the operation. In addition, syringomyelia is not detected. However, hyperintensity on T2-weighted imaging consistent with myelomalacia persisted after cyst resection (Figure. 2 A, B, C).

According to ENMG in the postoperative period, there were positive dynamics in the parameters of the speed of the radicular nerves in the L1-2 segments and the amplitudes of the M-response in the femoral and tibial muscles from 2 sides. The speed of conduction along the efferent fibers approached normal age parameters, which indicated the restoration of spinal conduction disorders from the level of the thoracic region due to the resolution of syringomyelia. Ultrasound of the kidneys 20 days after surgery showed reduced residual urine after urination. Physiotherapy and rehabilitation were prescribed 1 month after the operation. After 2 months, no neurological deficit was detected at the follow-up examination. In addition, a significant reduction in myelomalacia on MRI was observed after 2 months (Image 3D).

Discussion

Epidermoid cysts of the spine are acquired or congenital. Acquired epidermoid cysts described in the literature are due to iatrogenic penetration of skin fragments, often after a lumbar puncture or after surgery to remove the meningomyelocele (1, 8, 9). A congenital epidermoid cyst of the spine forms when ectodermal tissues are turned on during neural tube closure between the third and fourth weeks of embryonic development (3). Various spinal dysraphisms, *spina bifida aperta*, *hemivertebra*, and dermoids are often associated with congenital cysts (1, 3, 8). Our patient did not have a meningomyelocele, nor any trauma or disease leading to a lumbar puncture. Therefore, the tumor was considered congenital. Signs and

symptoms due to the presence of epidermoid cysts vary depending on the degree of involvement and do not differ from other lesions occupying the space of the spinal canal. Paraparesis, sensory disturbances, urologic manifestations, abnormal reflexes, profound changes in tendon reflexes, and pain in the back and extremities are usually found on examination. The slow growth of epidermoid cysts often results in late diagnosis (2).

MRI is important for diagnosis, especially in studies with contrast, which accurately shows the location and size of the lesion. MRI clarifies that the epidermoid cyst is hypointense on T1-weighted and hyperintense on T2-weighted images (8). Gadolinium enhancement improves diagnosis. During the operation, we found a dense capsule of the cyst. Electroneurophysiological examination makes it possible to accurately determine the level of predominant damage to spinal structures in multilevel pathological processes, indicating the predominance of a functional deficit at the segmental or conductor level. In particular, compression of the spinal structures by the tumor at the L1-2 level caused segmental disorders. Syringomyelia at the level of the thoracic region was manifested by moderate conduction disorders according to ENMG data and was secondary. Ependymomas are the most common tumors occupying space at the level of the conus of the spinal cord and the cauda equina. Lymphomas, astrocytomas, gangliogliomas, and ganglioneuromas may be considered in the differential diagnosis (7, 10, 11). They can often be identified and distinguished on MRI examinations. Astrocytomas, gangliogliomas, and ganglioneuromas have heterogeneous contrast features. Ependymomas have uniform enhancement, while epidermoid cysts have peripheral enhancement (7, 9, 11). However, radiological differentiation is often difficult for these spinal cord tumors.

Histopathological examination is critical for the diagnosis of epidermoid cysts. The difference between the two types of cystic tumors (dermoid and epidermoid) is based on the presence or absence of skin appendage structures such as sebaceous and sweat glands, hair follicles. They are present only in dermoid cysts (1, 2). The specific histological feature of an epidermoid cyst is a flattened keratinized stratified squamous epithelium surrounded by an outer layer of collagenous tissue and/or inflammatory cells without dermal appendages. Desquamation of keratin from epithelial tissue generates cholesterol crystals (2, 9).

Complete removal of the encapsulated tumor is a radical treatment for an epidermoid cyst of the spinal cord because it is a benign lesion. Sometimes, due to its anatomical features, an intramedullary tumor coarsely fuses with the arachnoid membrane and spinal cord, thereby causing difficulties for safe resection. Consequently, subtotal resection is often performed to avoid neurological deficits (1, 2). Local recurrence is more often noted after subtotal resection - incomplete removal of the basal germ cells of the tumor is the cause of relapses (12). Neurosurgeons should remember that cysts contain fat and cholesterol. This can trigger an inflammatory reaction that leads to aseptic chemical meningitis (Mollaret’s meningitis) (8). Plugging the proximal and distal arachnoid spaces and irrigating the spinal cord and nerve roots with warm saline before dura closure reduces the risk of chemical meningitis (9). We performed a complete removal of the epidermoid cyst,

so the patient did not develop any inflammatory reaction and neurological disorders in the postoperative period. The exact pathogenesis of syringomyelia associated with spinal cord tumors is unclear. Several different theories have been proposed. Some of them have an intramedullary explanation, due to circulatory disorders, CSF stagnation due to obstruction of the drainage pathways, edema, and spontaneous autolytic tumor reactions (7,13). Another possible mechanism for the development of syringomyelia is the blockage of cerebrospinal fluid (13, 10, 14). This explains why syringomyelia is often seen in intramedullary tumors. However, extramedullary tumors associated with syringomyelia are very rare.

In our case, syringomyelia most likely developed a second time due to the pressure of the epidermoid cyst on the cone of the spinal cord. We believe that the cyst caused the CSF flow block. Therefore, the removal of the cyst eliminated the mass effect and restored these changes in CSF flow. Thus, complete resection of the cystic tumor led to the disappearance of syringomyelia and eliminated the need for an additional procedure for draining the syringomyelia cavity.

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