MRI of an Endodermal Cyst of Cervicothoracic Spine: a case report

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An 8-month-old female baby presenting with neck stiffness was found to have an intradural extramedullary cystic mass at C3 to T2 level on magnetic resonance images (MRI). The spinal cord posterior to the mass was compressed. Surgery and pathology revealed endodermal cyst. Repeated surgery was performed 14 months after the first operation because of local recurrence. The 27 months of clinical course after the second surgery was uneventful.

Key words: Endodermal cyst; Spinal cord; Magnetic resonance imaging

An endodermal cyst, also called enterogenous cyst or neurenteric cyst, is a rare condition affecting the spinal cord. The clinical manifestations vary from spinal cord compression to neurological deficit. The pathology is a cyst lined by mucin-secreting epithelium which resembles that of the gastrointestinal tract. Magnetic resonance imaging (MRI) is valuable for such lesions in determining its location, extent, and content. Surgical removal is the choice of treatment and it rarely recurs if the attachment to the spinal cord is completely cleared.

MRI of an 8-month-old female baby with spinal endodermal cyst at the cervicothoracic level was presented. The pathogenesis and prognosis of such a lesion is discussed.

CASE REPORT

An 8-month-old female baby was noted to have neck stiffness and difficulty in turning her head to the right side for 1 month before admission. She was born full-term by a healthy mother. The prenatal course was uneventful. After admission, her general condition was well except for neck stiffness with inability to turn to the right side. No neurologic deficit was found. Laboratory examinations including blood chemistry and differential WBC count were within normal limit.

MRI of the cervicothoracic spine was performed on a 1.5T system. Axial as well as sagittal T1-weighted images, T2-weighted images, and T1-weighted images after Gd-DTPA enhancement were obtained. There was a large cystic lesion with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images between the vertebral bodies and the spinal cord from C3 to T2 level. It appeared intradural and extramedullary because of sharp angle both between the spinal cord and the vertebral bodies. The regional spinal cord was flattened due to long term compression (Fig. 1). After intravenous administration of Gd-DTPA, there was no enhancement in the lesion (Fig. 1a).

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Under the impression of an intradural extramedullary cystic lesion, laminectomy from the C3 to T2 level with excision of the tumor was carried out.

Surgery confirmed the intradural extramedullary location of the cyst. Pathology showed that the inner wall of the cyst was lined by a simple columnar epithelium with underlying fibrous stroma. An endodermal cyst or enterogenous cyst was diagnosed by the presence of simple columnar epithelium.

The patient was discharged with improvement in mobility of the neck and she was regularly followed up at the pediatric outpatient service.

Recurrence of the endodermal cyst at C5 to T1 level was found by MRI two months after the operation. An anterior intradural extramedullary cyst with mild compression over the spinal cord was disclosed (Fig. 2). A follow-up MRI 14 months later revealed progressive enlargement of the cystic lesion. The regional spinal cord was compressed, posteriorly displaced and flattened with concave anterior surface (Fig. 3). A second surgery was performed. The pathology proved the lesion to be a leptomeningeal endodermal cyst. The patient was discharged uneventfully and there was no clinical evidence of recurrence 27 months after the second operation.

**DISCUSSION**

An endodermal cyst is a cyst lined by mucin-secreting epithelium, resembling that of the gastrointestinal tract [1]. The condition is rare. Possible etiology proposed by most authors [2-9] is that it arises during the development of the notochord. The noto-
chord and the foregut may fail to separate completely in the third week of gestational age during formation of the definite alimentary canal. Persistence of the transient neurenteric canal may interfere with notochord development, as endodermal elements herniate through the cleft. The notochord halves later unite and sequester the intraspinal endoderm that can give rise to an endodermal cyst.

There are three histologic groups of neurenteric cysts or endodermal cysts [8,10,11]. Group A, or type I is the simplest, lined by ciliated or nonciliated epithelium supported by connective tissue. Group B or type II cyst has other elements along the tracheobronchial or gastrointestinal tracts in addition to the epithelial lining, these include mucus or serous glands, smooth muscle, fat and cartilage. In group C or type III cysts, there are co-existence of ependymal and glial tissues. Most group A cysts are located anterior to the spinal cord and those posterior to the spinal cord are located at the level of the conus medullaris or cauda equina [2]. However, most of the group B and C cysts are posterior to the cord regardless of the position along the rostral-caudal axis. The cyst with simple columnar epithelium in this patient was located anterior to the cord at the cervicothoracic level, a finding consistent with the group A cyst.

About half of the reported cases were associated with congenital malformations such as spina bifida, fused vertebrae (block vertebrae), hemivertebrae, or anterior spina bifida [12,13]. This patient had no such associated malformations.

MRI is currently the best imaging modality to detect endodermal cysts partly due to its noninvasive-ness [2]. It can clearly demonstrate the exact location and extent of lesions, and differentiate the tissue characteristics by different pulse sequences and intravenous administration of Gd-DTPA. In this case, the lesion was apparently intradural extramedullary in location from the C3 to T2 level. With the hypointensity on T1-weighted images and non-enhancement on Gd-enhanced T1-weighted MR images, it is easy to determine its cystic nature.

The differential diagnoses of intradural cystic masses include epidermoid cyst, dermoid cyst, arachnoid cyst, myelomalacia, intradural or intramedullary abscess, syrinx and endodermal cyst [14]. Epidermoid cyst can be either congenital or acquired. In this case, the absence of epidermal defect such as spina bifida or hemivertebrae might exclude the possibility of congenital epidermoid cyst. Furthermore, the denied history of spinal trauma or lumbar puncture excluded the possibility of acquired epidermoid cyst. Non-delineation of the fat signal on MRI in this case might exclude the possibility of a spinal dermoid, which is uncommon and constitutes 20% of intradural tumors seen during the first year of life. Intradural arachnoid cysts are rare, mostly communicate with subarachnoid space and opacify following intrathecal contrast administration. This baby had no fever and leukocytosis, nor peripheral enhancement on MRI, thus spinal abscess could be excluded. The diagnosis of syrinx also was unlikely due to the extramedullary location of the lesion.

The treatment of choice for endodermal cyst is surgery. Due to the tight adhesion of the lesion to the spinal cord, as shown in this patient, complete excision is sometimes difficult [15]. Partial removal of an endodermal cyst may carry risk of recurrence, although reported cases of recurrence and metastasis after surgery are extremely rare [13]. As was shown in this case, surgical removal was the only choice of treatment to relieve the symptoms in cases of local recurrence.

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頸胸椎內皮囊腫之磁共振造影：病例報告

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一個八個月大的女嬰因頸部僵硬就診。磁共振造影顯示由第三節頸椎至第二節胸椎的脊髓腔內有一硬膜內細外的囊狀病灶，此病灶將脊髓往後壓扁。手術及病理顯示為內皮囊腫。因為局部復發，此女嬰在十四個月後再度接受手術。第二次術後的二十個月追蹤期內並無復發跡象。

關鍵詞：內皮囊腫、脊髓、磁共振造影