

Case Report

Bronchogenic cyst of the conus medullaris with spinal cord tethering: a case report and review of the literature

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Received February 2, 2015; Accepted March 30, 2015; Epub April 1, 2015; Published April 15, 2015

Abstract: Bronchogenic cysts (BCs) are congenital malformations that originate from remnants of the primitive foregut. Intraspinial BCs, especially those of the conus medullaris are rare with only one case reported until now. To date, a bronchogenic cyst with spinal cord tethering has not been previously reported. We reviewed the clinical course of a 44-year-old woman, who presented with low back pain and leg weakness as well as sphincter disturbance. Magnetic resonance imaging showed an intradural oval mass located at the conus medullaris. A tethered cord was also observed, as well as a dermal sinus tract. The mass was totally removed after an L3-L4 laminectomy without detethering during operation. Pathologic examination confirmed the diagnosis of bronchogenic cyst. By six months after treatment, the patient had experienced nearly complete recovery. The review of literature indicated that detethering was performed in most reported cases of neurenteric cysts with spinal cord tethering, and one of six patients was diagnosed with a postoperative recurrence. The co-existence of bronchogenic cyst and a tethered spinal cord would imply associated developmental errors in embryogenesis. It is worth noting that whether detethering is necessary after the cyst removal.

Keywords: Bronchogenic cyst, conus medullaris, tethered spinal cord, intraspinal cyst, neurenteric cyst

Introduction

Arising from abnormal budding of the primitive foregut, bronchogenic cysts are benign malformations and often classified as one of histopathological subtypes of neurenteric cysts. According to the study of Zambudio et al. [1], bronchogenic cyst was the most common non-neoplastic mediastinal cyst, accounting for 50-60% of this pathology. Although prior studies have reported cases of these cysts occurring in cutaneous and subcutaneous tissues, neck, pericardium, diaphragm and abdomen [2], intraspinal cases, especially those with the involvement of the conus medullaris, are rarely reported. In 2009, Yilmaz et al. [3] reported a case of intramedullary bronchogenic cyst of the conus medullaris and reviewed 8 cases of spinal bronchogenic cysts in eight publications. Since then, an additional two such cases have been documented in the English literature [4, 5]. To the best of our knowledge, however, a localized bronchogenic cyst of the conus medul-

laris with spinal cord tethering has not been previously reported to date. We describe here a unique case and discuss this co-existence in the context of embryogenesis with a literature review.

Case report

History and examination

A previously healthy 44-year-old woman presented with a 9-year history of low back pain and 2-year-long weakness in lower extremities, which gradually worsened in the latest 2 months. On admission, she had moderate sphincter disturbance with indwelling catheter. Physical examination revealed light tenderness and cutaneous pigmentation with a diameter of 4.0 cm over the lumbosacral region. Neurological examination showed decreased muscle strength of both lower extremities (grade 4 of 5) and negative bilateral knee and ankle reflex. A mild hypoesthesia in sellar area was also observed. Magnetic resonance imaging

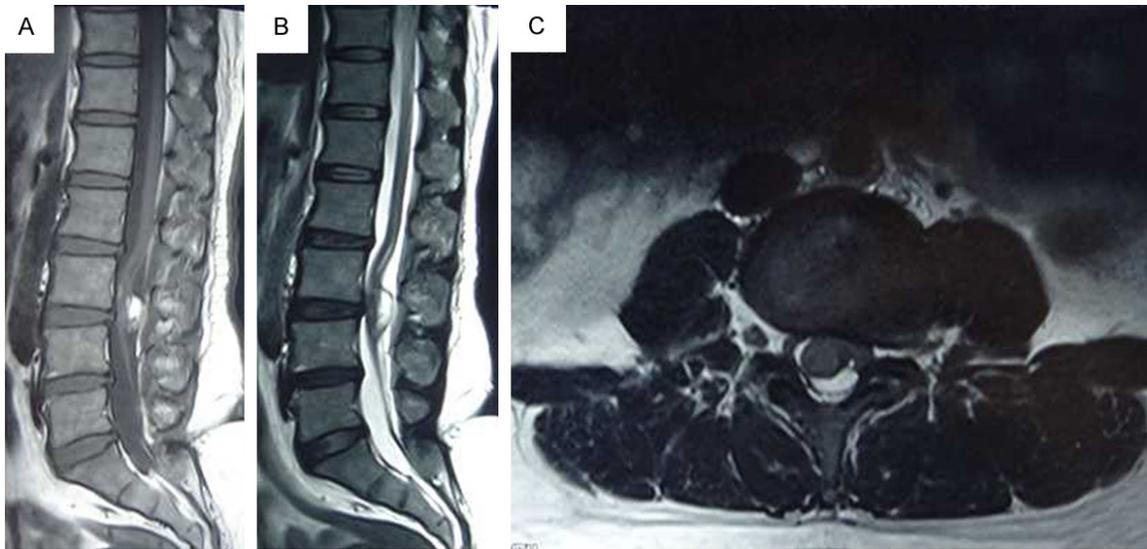


Figure 1. Sagittal T1-weighted (A), sagittal T2-weighted (B), and axial T2-weighted (C) MR images showed an intradural mass located in the dorsal part of conus medullaris. There was also a tethered cord that was low lying and terminated at the level of L4, as well as a dermal sinus tract at the S1 level.

(MRI) of the lumbosacral spine showed a well-circumscribed, 2.0×1.5×1.0 cm intradural oval mass located at the conus medullaris. The lesion had a mixed hyperintense and isointense compared to cord on T1- and was hyperintense on T2-weighted images. There was also a tethered cord that was low lying and terminated at the level of L4, as well as a dermal sinus tract at the S1 level (**Figure 1A-C**).

Operation

The patient underwent an L3-L4 laminectomy. A 4 cm midline incision was made to the thecal sac. A severe compression of the conus medullaris caused by a mass was encountered. Although no adhesion to the thecal sac was observed, the base of the mass was tightly attached to the conus and cauda equina. The adhesions were meticulously separated, and then the mass was totally removed. No detethering of the spinal cord was performed during surgery.

Histopathological examination

The dissected mass revealed cystic and filled with yellow viscous fluid. Histological examination of the specimen with hematoxylin-eosin (H&E) staining showed the cyst wall was lined by simple and pseudostratified respiratory epithelium with benign subepithelial mucous glands and fat components neighboring the

cyst (**Figure 2A, 2B**). Histopathological findings were consistent with bronchogenic cyst.

Postoperative course

The patient's symptoms improved gradually after surgery. She was discharged on the 18th postoperative day. At the six-month follow-up study, no symptoms or signs of cyst recurrence were observed. The patient denied disturbance of bladder and bowel, and also had no complaint of back pain as well as weakness in both legs. She was continued to be seen for follow-up visits for at least one year.

Discussion

Neurenteric cysts, also called enterogenous cysts, endodermal cysts, or foregut cysts, are anatomical malformations that originate from congenital remnants of primitive foregut as a result of the abnormal partitioning of presumptive endoderm and the embryonic notochordal plate during Day 21 of embryogenesis [5]. These congenital lesions are relatively uncommon and account for only 0.7-1.3% of all spinal cord tumors [6]. On histopathological analysis, neurenteric cyst is called a bronchogenic cyst if it has lining epithelium consistent with that of the respiratory tract. Previous studies have shown that bronchogenic cyst could be found attached to the sternum, skin, abdomen, pericardium and diaphragm [2], but rare cases of

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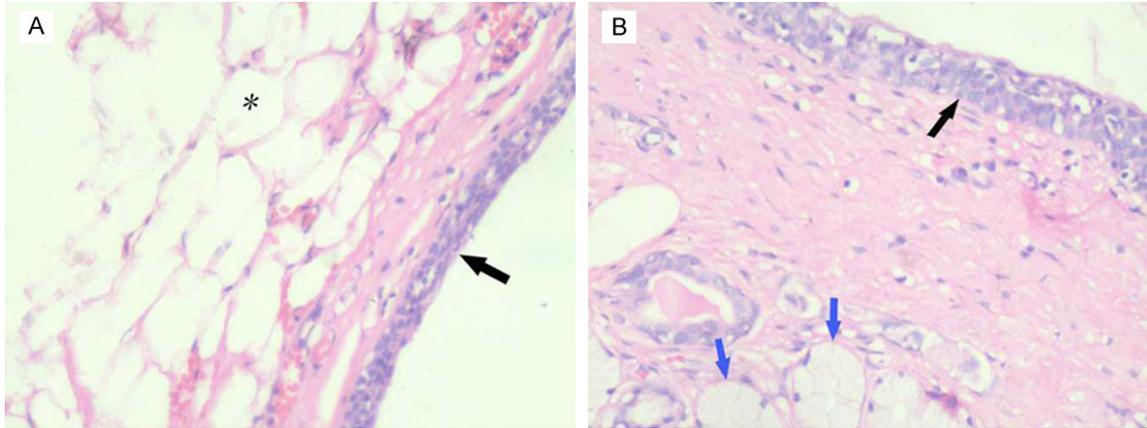


Figure 2. Photomicrographs showed the cyst was lined by (A, B) simple and pseudostratified respiratory epithelium (black arrow), (A) with normal fat components neighboring the cyst (asterisk). (B). The benign subepithelial mucous glands neighboring the cyst was also observed (blue arrow). H&E, original magnification $\times 200$.

intraspinal involvement have been also reported [3]. According to the Wilkins and Odom neuroenteric cyst histopathological classification system [7], neuroenteric cysts have three types: type A cysts contain columnar or cuboidal cells, with ciliated and nonciliated components; type B cysts contain all features for type A and have some additional tissues, including lymphatic tissue, bone, cartilage, fat or glandular components; type C cysts are diagnosed by ependymal or glial tissue and type A features. In the case presented here, the cyst was classified into type B because the histopathological examination appeared respiratory epithelium, mucous glands and fat components.

MRI is the first radiologic modality of choice for making an early diagnosis because of the advantage in delineating the cystic lesion and evaluating its relationship with surrounding neural structures. Currently, there is lack of summary of the MRI characteristics for spinal bronchogenic cyst. Liu et al. [5] has suggested that the most common MRI findings for this disease are noncontrast-enhancing lesions that are hypointense on T1- and hyperintense on T2-weighted imaging. However, our case seemed to present with an increased signal intensity on both T1- and T2-weighted images, which might provoke a strong clinical concern for intradural lipoma initially as lipomatous tumors were frequently seen, along with a tethered spinal cord [8-10]. In fact, signal change for the cyst was heterogeneous on the initial MRI, presenting as a mixed hyperintense and isointense compared to the cord. Furthermore,

subsequent histopathological examination confirmed the diagnosis of bronchogenic cyst as opposed to lipoma, although normal fat components were also observed. This finding suggests that frequent variations may be seen for MRI findings associated with these cysts, consistent with previous reports [11].

One unique presentation of our case was that the cyst developed concurrently with a tethered spinal cord. Past reports on the co-existence of neuroenteric cysts and spinal cord tethering in adults has been rarely described in the English language literature, with only six cases documented to date [12-17]. **Table 1** summarized previous studies of intraspinal neuroenteric cysts with a tethered cord in adults. In these cases, 3 arose in the lumbar region, 1 in the thoracic region, and 2 in the thoracolumbar region. However, no cases had highlighted the type of lesion of bronchogenic cyst.

To our knowledge, tethered cord syndrome represents various developmental intraspinal anomalies that bind the distal spinal cord low in the bony spinal canal [9]. This entity is uncommon in adults, with bowel and bladder dysfunctions most frequently encountered [9, 18]. Based on these characteristics, some authors may have a concern that the sphincter disturbance presented in our patient could be likely caused by the initially diagnosed tethered spinal cord. Previous studies have identified several factors contributing to the onset of symptoms of adult tethered cord syndrome, including an increase in physical activity, develop-

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Table 1. Summary of previous studies of intraspinal neurenteric cysts with spinal cord tethering in adults

Authors, Year	Age (y)	Sex	Clinical features	Bladder and rectal disturbance	Lesion Location	Type of Lesion	Result of biopsy	Extent of Resection	Follow-Up	Detethering	Complication	Outcome
Bakaris et al., 2005 [12]	31	M	Low back and lower limbs pain	-	L4-L5	Intradural extramedullary	Neurenteric cyst	Partial	NA	Yes	No	Completely symptom-free
Freund et al., 1998 [13]	60	M	Low back and leg pain, paralysis of the left leg	-	L2-L3	Intradural extramedullary	Neurenteric cyst	Total	NA	NA	NA	NA
Rauzzino et al., 2001 [14]	45	F	Progressive paraparesis	+	T4-T7	Intradural extramedullary	Neurenteric cyst	Total	NA	NA	No	Leg weakness and bladder dysfunction improved but not normalized, recurrence
Menezes et al., 2006 [15]	55	M	Back pain and episodic weak legs, small right foot since birth, decreased sensation to touch and pinprick in the left leg	-	T12-L1	Intradural extramedullary	Neurenteric cyst	Total	12 yr	Yes	NA	Leg weakness resolved as well as pain, no recurrence
Vachhani et al., 2012 [17]	35	F	Progressive weakness and loss of coordination in her legs	+	T12-L1	Intramedullary	Neurenteric cyst	Partial	NA	Yes	NA	Strength and coordination improved
Jain et al., 2003 [16]	34	M	Burning sensation in both legs	-	L2-L5	Intradural extramedullary	Neurenteric cyst	Total	NA	Yes	No	Completely symptom-free

ment of spinal stenosis, trauma to the lumbar spine and straight leg exercises [10, 19-21]. However, these predisposing factors are rarely present in our patient. We postulate that the bronchogenic cyst as a slow growing lesion may lead to the patient's symptoms by a chronic compression. Alternatively, the asymptomatic tethered spinal cord in this patient was pushed over the threshold from her progressively growing cystic lesion, thereby resulting in the onset of symptoms. Considering these speculations, detethering of the spinal cord was not performed during operation, although it was made in most of the reported cases of neurenteric cysts with a tethered spinal cord [12, 15-17] (**Table 1**). However, the choice of our strategy can be further justified by the satisfactory outcome in her subsequent follow-up.

Bronchogenic cyst accompanied with spinal cord tethering in the present case suggests that dysembryogenesis is likely involved in the development of this co-existence. Although the exact mechanism for the development of bronchogenic cysts remains unclear, one widely accepted hypothesis is the split notochord syndrome [22]. It states that partial duplication and separation of the notochord may develop in an embryo, resulting in a gap formation between the two parts of the notochord, through which the ventrally situated yolk sac or gut anlage endoderm may herniate. Further, the hernia may rupture to produce a fistula between the yolk sac and the amniotic cavity that divides the future cord and spine. Subsequent differentiated growth of the embryo may close the fistula and lead to a cystic lesion. Given the time and location of the embryologic failure [23], we speculate that some common developmental errors should be implicated in the comorbidity of our case. Furthermore, in agreement with Takci et al. [24], we also believe that the congenital tethered spinal cord may have caused the development of the cyst.

Although the prognosis for this cyst is excellent [25], rare case of postoperative recurrence has been reported [26] and retethering of the spinal cord may occur [8], which necessitates close follow-up especially when signs of deterioration or discontinuation of improvement appear. In fact, we are planning to conduct follow-up examinations in our patient periodically for at least one year.

In conclusion, our case represents a very rare condition. Although surgery seems to be an encouraging treatment in these patients, aggressive neurosurgical intervention for the treatment of coexisting spinal cord tethering needs due consideration. However, postoperative radiologic follow-up is advised especially when signs of deterioration or cessation of improvement appear, in an attempt to detect clinically undesirable events and implement appropriate treatment, if necessary.

Disclosure of conflict of interest

None.

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