Case Report

An unusual case of intradural intramedullary dorsal bronchogenic cyst in spine

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Abstract: Bronchogenic cysts are congenital malformations derived from anomalous budding of the embryonic foregut. Intraspinal bronchogenic cysts are extremely rare and most of them are extramedullary. There has been only one case of intramedullary spinal bronchogenic cyst reported. We present an 18-year-old male patient with spastic paraparesis and bowel and bladder involvement. MRI revealed a 2 cm diameter intradural and intramedullary lesion at D2–D3 level which was hyperintense on T1 and hypointense on T2 imaging. Histopathological examination after surgical excision of the lesion revealed a bronchogenic cyst. To our knowledge this is the first case reporting an intramedullary bronchogenic cyst at the upper dorsal level and overall second reported case of intramedullary spinal bronchogenic cyst.

Keywords: Upper dorsal; bronchogenic cyst; intradural; intramedullary

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Introduction

Bronchogenic cysts are congenital malformations derived from anomalous budding of the embryonic foregut. It is considered the 2nd most common foregut duplication cyst after neurenteric cysts (1). These are also considered as the most common non-neoplastic mediastinal cysts (2). These are usually solitary, but multiple may be found in a patient and can be filled with fluid or proteinaceous material. These have been also reported in more remote locations like the neck, abdomen and retroperitoneal space. Intraspinal bronchogenic cysts are extremely rare and most of them are extramedullary (3,4). Most of them are located in the cervical or upper thoracic region. To the best of our knowledge, there has been only one case of intramedullary spinal bronchogenic cyst reported (5). We therefore present a unique case of intradural intramedullary spinal bronchogenic cyst.

Case presentation

History and examination

An 18-year-old male presented with complaints of upper back pain and numbness and weakness in both of his lower limbs. He explained that he was able to ambulate only with support for the last three years. The weakness of lower limbs was gradually progressive and now he has not been walking even with support for the last two months. There was difficulty in voiding his bladder and evacuating his bowel for the last two months. The patient was otherwise medically fit.

On clinical examination, the patient had bilateral spastic lower limbs with muscle wasting and unable to move against gravity. Sensations were decreased below the nipples. His knee and ankle jerks were brisk and abdominal reflex was absent. Babinski sign was positive.

Magnetic resonance imaging (MRI) of the patient's

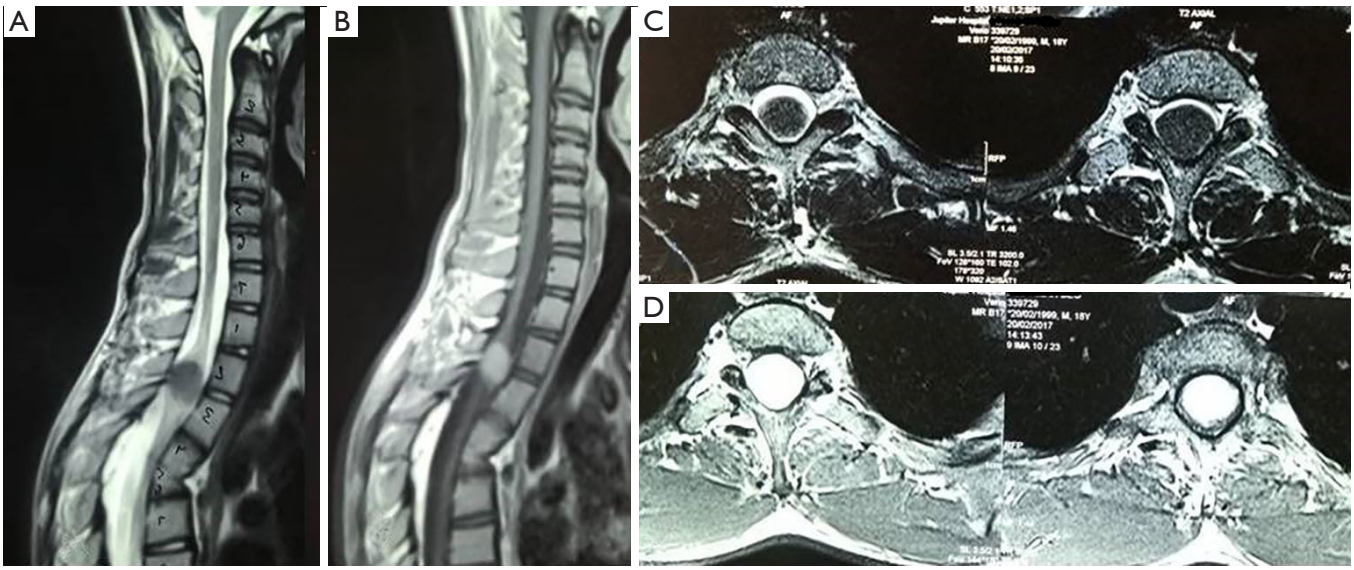

Figure 1 Magnetic resonance imaging of the lesion showing intradural and intramedullary location. (A) T2 weighted sagittal image in which lesion is hypointense; (B) T1 weighted sagittal image in which lesion is hyperintense; (C) T2 weighted axial view; (D) T1 weighted axial view.

spine showed there to be an intradural intramedullary lesion of 2 cm × 1.6 cm located at the D2–D3 level with fusion of D4, D5 and D6 vertebrae and localized kyphosis. The lesion was hyperintense on T1 and hypointense on T2 weighted images (*Figure 1*). It was causing significant compression of the spinal cord.

Operation

The patient underwent laminectomy at the D2–D3 level. A midline incision was made to the dura and cord. Yellowish viscous fluid was drained from the cyst (*Figure 2*). The cyst wall couldn't be resected completely because of adherence. Cystic fluid and tissue samples were sent for histopathological examination. Histopathological examination revealed strips of cyst wall partly lined with ciliated columnar epithelium and squamous epithelium (*Figure 3*). The collagenous wall showed focal lymphocytic infiltrate. There were loose aggregates of histiocytes with fibrinous material. Histopathological findings were consistent with a bronchogenic cyst.

Postoperative course

Patient showed some improvement in his neurology after one month post-operation. He is able to move his lower limbs against gravity. Patient was taught how to cleanly perform intermittent self-catheterization to void his bladder.

Discussion

Bronchogenic cysts are congenital abnormalities resulting from abnormal budding of the primitive foregut, identified as epithelial endodermal cyst. A lesion is termed as a bronchogenic cyst if endodermal lining is mostly pseudo stratified ciliated columnar epithelium (similar to respiratory tract mucosa).

The etiopathogenesis is not known but there are some hypothesis proposed, such as incomplete germ layer separation due to ecto-endodermal adhesions and the notochord syndrome theory. The notochord syndrome theory is widely accepted and is explained by Bantley and Smith (6). They described that partial duplication and separation of the notochord leads to a ventral herniation of the yolk sac or remnant gut endoderm through the notochord. Hence a fistula is formed with the amniotic cavity. As the embryo grows, the fistula closes and a cystic mass is formed from the remnant of foregut.

These lesions are considered slow growing in previous reports because of tight adhesions between epithelial

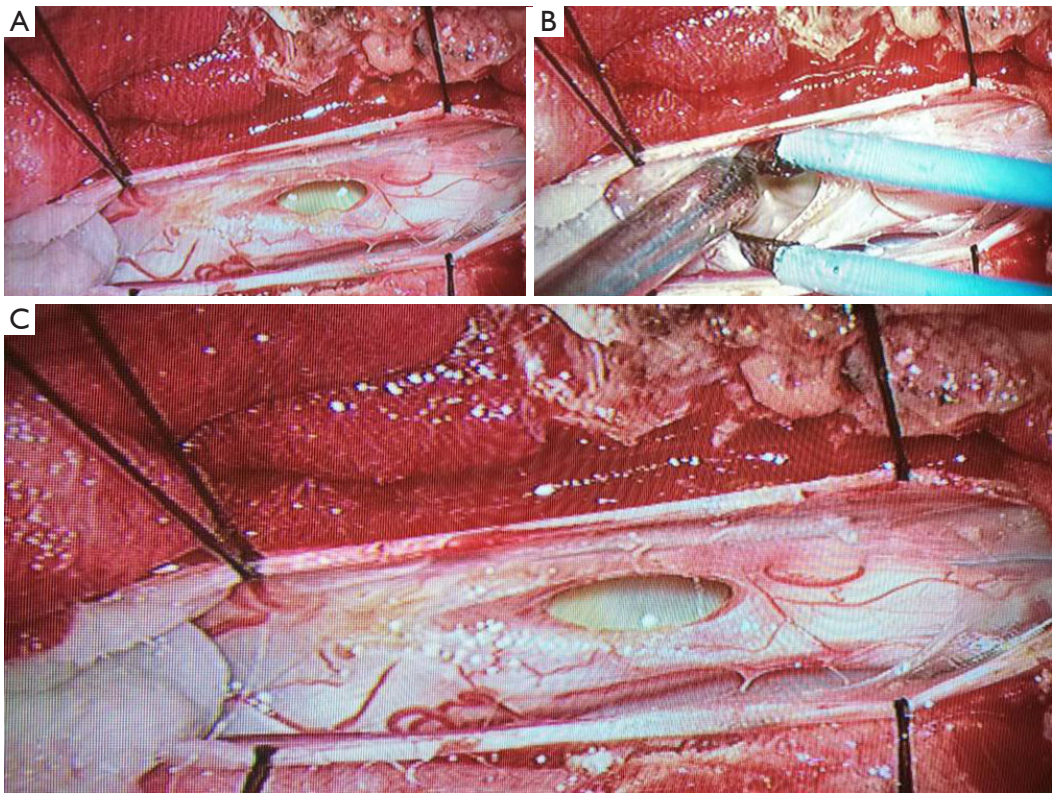

Figure 2 Intra-operative images. (A) Cystic material coming out automatically as the cord is opened; (B) drainage of cystic material; (C) after significant drainage of cyst.

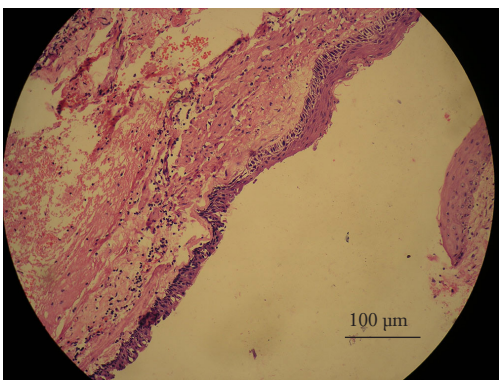

Figure 3 Histopathological picture of the collected specimen which is clearly showing pseudo-stratified columnar epithelium.

cells (6,7). Malignant transformation is only reported in mediastinal cysts, but not in spinal bronchogenic cysts. The most effective treatment is surgical resection but up to 11.6% recurrence is reported because of partial resection (8).

To the best of our knowledge, eleven cases have been reported in the literature (*Table 1*). Ten of them were intradural and extramedullary. Six of them were in the cervical or upper thoracic region, one in the lower thoracic, one in the thoracolumbar, two in the lumbar and one in the sacral region. To our knowledge, only one case of intradural and intramedullary spinal bronchogenic cyst has been reported as of yet.

In this case report, the patient developed symptoms during adolescence with severe weakness of his lower limbs and bowel and bladder involvement. The unique aspect of this case, apart from common features, is its intramedullary location. It was hypointense in T2 and hyperintense in T1 weighted imaging because of its high proteinaceous material content.

We conclude that spinal bronchogenic cysts are extremely rare with only 11 cases reported in literature. It can present with a vast range of symptoms starting from isolated pain to complete paraplegia. Surgical resection offers good results but recurrence is common. Therefore close follow-up is necessary after surgery.

Table 1 Review of literature: bronchogenic cyst in spine

Author's name, year	Symptoms	Lesion location	Type of lesion	Resection	Follow-up period
Yamashita *et al.* (9), 1973	Intermittent neck and left arm pain for 4 years	C6–C7	Intradural extramedullary	Total	11 months
Ho & Tiel (10), 1989	Numbness in right arm and leg for 6 weeks	C5–T2	Intradural extramedullary	Total	–
Wilkinson *et al.* (11), 1992	Pain, paraesthesia right arm for 2 weeks	C3–C4	Intradural extramedullary	Partial	1 year
Baba *et al.* (12), 1995	Suboccipital pain for 1 year	C1	Intradural extramedullary	Total	1 year
Rao *et al.* (13), 1999	Pain and progressive weakness right arm for 6 weeks	C2–C3	Intradural extramedullary	Total	3 months
Baumann *et al.* (14), 2005	Acute leg pain	T12–L1	Intradural extramedullary	Partial	3 Months
Chongyi *et al.* (3), 2008	Chronic lumbago for 1 year and progressive weakness and numbness of left leg for 2 weeks	L1	Intradural extramedullary	Partial	–
Ko *et al.* (15), 2008	Skin dimple at sacral region	S2	Intradural extramedullary	Total	9 days
Arnold *et al.* (8), 2009	Weakness and paraesthesia in lower limbs, urinary incontinence	T4	Intradural extramedullary	T4	1 year
Yilmaz *et al.* (5), 2009	Chronic back pain and lower limb paraesthesia for 2 months	T12	Intradural intramedullary	Partial	6 months
Viswapathi *et al.* (16), 2016	Buttock pain and numbness and right leg sciatica for 3 months	L3–L4	Intradural extramedullary	Total	–

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None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

Informed Consent: Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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