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

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References
11. Wilkinson N, Reid H, Hughes D. Intradural bronchogenic...