Thoracic Myelopathy Secondary to Intradural Extramedullary Bronchogenic Cyst

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Abstract

Background/Objective: To report a case of thoracic myelopathy secondary to intradural extramedullary bronchogenic cyst.

Study Design: Case report.

Methods/Findings: A 20-year-old man presented to the emergency department with increasing back pain and lower-extremity weakness. Magnetic resonance imaging demonstrated a cystic lesion at the T4 level with mass effect on the spinal cord.

Results: The lesion was resected, and histopathologic evaluation showed a cyst lined by respiratory-type epithelium consistent with a bronchogenic cyst.

Conclusions: Intradural extramedullary bronchogenic cysts of the thoracic spine have been reported previously but are extremely rare. The treatment of choice is surgical resection.

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Key Words: Bronchogenic cyst; Myelopathy, compressive; Neoplasm, extramedullary intradural spinal cord; Paraparesis

INTRODUCTION

Bronchogenic cysts are congenital anomalies that usually originate in the mediastinum; atypical locations include the sternum, pericardium, skin, diaphragm, and spinal cord (1). Intradural bronchogenic cysts of the spinal cord are rare and tend to occur most frequently in the cervical or upper thoracic region (1,2). Bronchogenic cysts enlarge slowly and may remain undetected for many years. Preferred treatment of symptomatic bronchogenic cysts is surgical removal; most persons experience symptomatic relief postoperatively.

CASE REPORT

A 20-year-old man presented with back pain of 6 months' duration. He also described the recent onset of urinary incontinence, numbness in the lower extremities, and increased difficulty in walking (he had required a wheelchair for 1 month prior to presentation). He had no significant past medical history.

Physical examination revealed 5/5 strength in the upper extremities with 4/5 in the quadriceps and plantar flexion. Sensation was intact to the level of approximately T6, with decreased sensation to light touch below that level. He had hyperreflexia in the lower extremities, with 3 beats of clonus bilaterally and Babinski's sign bilaterally. The patient was able to take several steps with assistance and displayed a wide-based gait. The rest of his examination was unremarkable.

Magnetic resonance imaging (MRI) of the spine showed an intradural extramedullary cystic lesion centered at the T4 level (Figure 1). The lesion was approximately 2 cm in diameter and compressed the spinal cord. MRI of the lumbar spine was normal. Because of the progressive myelopathic symptoms, surgical treatment was indicated. The patient underwent a T4 to T5 laminectomy. Intraoperative ultrasound demonstrated an intradural cyst dorsal to the spinal cord (Figure 2).

Intraoperatively, the cyst was identified as a discrete lesion and was dissected from the spinal cord. The dura was opened in the midline, and with the aid of an operating microscope, a large cystic mass was removed en bloc. The spinal cord was then inspected for other lesions; no other pathology was identified, and the dura was closed with a running suture. Histopathologic evaluation revealed a connective tissue base lined by ciliated columnar epithelium, consistent with a broncho-

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Figure 1. (A) Sagittal and (B) axial T2-weighted MRIs show a cystic lesion at T4. It fills the spinal canal and causes compression of the spinal cord.

genic cyst (Figure 3). The patient tolerated the procedure well and was discharged on the third postoperative day. His lower extremity weakness resolved, as did his urinary incontinence, although he remained hyperreflexic. There was no evidence of recurrence 1 year after surgery (Figure 4).

DISCUSSION

There are 3 types of intraspinal cysts: epithelial, mesenchymal, and mixed. Epithelial cysts lined by displaced epithelium, presumably from the embryologic primitive

Figure 2. Intraoperative ultrasound shows an echogenic lesion consistent with a cyst.

foregut (3), are termed neurenteric or enterogenous cysts. Neurenteric cysts are uncommon lesions and represent approximately 0.5% of spinal cord space-occupying lesions (4). Bronchogenic cysts are a type of neurenteric cyst lined by respiratory epithelium. The present cyst demonstrated ciliated columnar cells consistent with respiratory epithelium and was therefore classified as a bronchogenic cyst. The dorsal location of this thoracic-level cyst differs from several reports that cite the typical location of solitary neurenteric cysts as the ventral cervical spine region.

Bronchogenic cysts are developmental abnormalities that may affect the central nervous system. The pathogenesis is not completely understood, and 3 main hypotheses have been proposed. One theory suggests the possibility of an ecto-endodermal adhesion, which results in incomplete separation of germ cell layers (5). A second theory suggests that the cyst originates from ectoderm, which is capable of producing both endodermal and mesodermal tissues (5). A third theory, the split notochord syndrome postulated by Bentley and Smith (6), describes an incomplete duplication and separation of the notochord that leads to a ventral herniation through the notochord, resulting in a fistula. This fistula runs through the future spinal cord and spine and is eventually closed off as the embryo grows, but a cystic lesion may remain on the ventral surface. The cyst is therefore a remnant of the foregut, so it can differentiate into tissues of the gut or any of the endodermal derivatives. Vertebral anomalies, more commonly seen at the lumbosacral level, are often associated with the split notochord syndrome but were not noted in this patient.

Bronchogenic cysts grow slowly owing to tight junctions between epithelial cells, limiting expansion of the cyst. The most effective treatment of a symptomatic spinal bronchogenic cyst is surgical resection; however, complete resection can be challenging if the cyst is adherent to the spinal cord. Only 36% of cysts reported are completely removed; the remainder are adherent (7). There are no reports in the literature of

Figure 3 (A) The resected cyst is composed of a fibrous wall lined by columnar, often pseudostratified, epithelium with areas of cilia present (arrow and inset) (hematoxylin and eosin, original magnification $\times 16$; inset, $\times 40$) (B). Cyst epithelium shows strong immunoreactivity for cytokeratin antisera. An identical staining pattern was seen with cytokeratin 7 antisera, but no immunoreactivity was observed with cytokeratin 20, a profile typical of respiratory tract epithelium (antisera to pan-cytokeratin, original magnification $\times 11.5$).

recurrence after total removal of the lesion (8). The overall recurrence rate reported in the literature is 11.6%, which is attributed to partial resection of the cyst in all cases (9).

CONCLUSION

Spinal intradural extramedullary cysts are a rare cause of symptomatic spinal cord compression (10) and should be completely resected whenever possible to prevent recurrence.

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Figure 4. Postoperative MRI reveals no evidence of the cyst.

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