Intramedullary bronchogenic cyst in the foramen magnum region accompanied with syringomyelia
A case report and literature review

Fan Chen, MDa,b, Sascha Marx, MDb, Chaochao Zhang, MDa, Junguo Cao, MDa, Ying Yu, MDa, Dawei Chen, MDa,*

Abstract
Rationale: Bronchogenic cysts refer to congenital anomalies derived from the primitive foregut. Spinal bronchogenic cysts are uncommon entities, and those occurring in the intramedullary sites are extremely rare. Bronchogenic cysts involving the foramen magnum region have only been described in 2 cases; however, intramedullary bronchogenic cysts with syringomyelia have not yet been reported.

Patient concerns: A 46-year-old woman presented with a 6-month history of pain in the posterior neck region and a 1-month history of numbness in the upper extremities. Neurological examination revealed a loss of sensation in bilateral upper extremities and sensory dissociation. Magnetic resonance imaging (MRI) showed an intramedullary cystic lesion in the foramen magnum region and syringomyelia.

Diagnosis: Histopathological findings were consistent with a bronchogenic cyst.

Interventions and outcomes: A surgical resection of the cystic lesion was performed via a posterior midline approach. Under neurophysiological monitoring, the cyst was punctured, yielding gelatinous liquid. The dorsal part of the cystic wall was removed. One month postoperatively, the symptoms were resolved completely. Three months after operation, MRI showed no recurrence of the cyst and the syringomyelia disappeared.

Lessons: Intramedullary bronchogenic cysts with syringomyelia are extremely rare. Preoperative identification is challenging and definitive diagnosis depends on histopathological evidence. Timely surgical resection should be highlighted.

Abbreviations: C = cervical, CT = computed tomography, EMA = epithelial membrane antigen, F = female, Gd-DTPA = gadolinium-diethylenetriamine penta acetic acid, GFAP = glial fibrillary acidic protein, GTR = gross total resection, L = lumbar, M = male, MRI = Magnetic resonance imaging, STR = subtotal resection, T = thoracic.

Keywords: bronchogenic cyst, case report, intramedullary, surgical resection, syringomyelia

1. Introduction
Bronchogenic cysts refer to congenital anomalies derived from the endoderm of the developing respiratory system. Pathologically, this entity is typically lined with pseudostratified ciliated columnar epithelium. Bronchogenic cysts are more frequent in paediatric patients, and they are frequently found in the mediastinum followed by digestive tract, pericardium and skin.[1] Spinal bronchogenic cysts are uncommon entities; especially, those occurring in the intramedullary sites are extremely rare. Due to the rarity of spinal bronchogenic cysts, the origin of these entities has not been fully understood, and the clinical and radiological characteristics as well as the treatment are not well known. In previous literatures, bronchogenic cysts involving the foramen magnum region have only been described in 2 cases[2,3]; however, intramedullary bronchogenic cysts with syringomyelia have not yet been reported.

Herein, we reported a case with intramedullary bronchogenic cysts in the foramen magnum region and accompanying syringomyelia. The clinical, radiological and histopathological profiles were analyzed, and relevant literatures were reviewed.

2. Case report
2.1. History and examinations
A 46-year-old woman presented with a 6-month history of pain in the posterior neck region and a 1-month history of numbness in the upper extremities. Neurological examination revealed a loss of sensation in bilateral upper extremities and sensory dissociation. The MRI showed an intramedullary cystic lesion in the foramen magnum region with accompanying syringomyelia.
The lesion was hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging; after gadolinium-diethylenetriamine pentaacetic acid (Gd-DTPA) administration, the cystic wall showed heterogeneous enhancement (Fig. 1). A preliminary diagnosis of intraspinal cyst in the foramen magnum region was made.

2.2. Surgery and pathology

A surgical resection of the cystic lesion was performed via a posterior midline approach. Partial occipital bone, atlas arch, and axis arch was removed. The dural mater was incised, and no spinal cord pulsation was observed. Under neurophysiological monitoring, the cyst was punctured, yielding gelatinous liquid. The dorsal part of the cystic wall was removed. The cystic wall was greyish-white and translucent with a thickness of 1.5 mm. The spinal cord pulsation was recovered. Intraoperative neurophysiological monitoring displayed no loss of somatosensory or motor evoked potentials.

Histopathological examination of the resected cystic wall showed pseudostratified ciliated columnar epithelium with abundant cilia, which were consistent with a bronchogenic cyst (Fig. 2). No immunohistochemical staining was performed.

2.3. Postoperative course

The postoperative course was uneventful, and the neck pain and upper-extremity numbness were relieved immediately. One month postoperatively, the symptoms completely resolved. Three months after operation, MRI showed no recurrence of the cyst and the syringomyelia disappeared (Fig. 3).

3. Literature review

In literatures, a total of 20 cases with spinal bronchogenic cyst were identified, including 11 males and 8 females. The ages ranged from 5 months to 66 years (mean 31.9 ± 16.8 years). The clinical manifestations of spinal bronchogenic cysts were nonspecific, including local pain and extremity sensorimotor deficiencies. On MRI, spinal bronchogenic cysts manifested as isointensity (3/11) or hypointensity (8/11) on T1-weighted imaging, and hyperintensity (16/16) on T2-weighted imaging; after Gd-DTPA administration, slight (2/8) or no (6/8) enhancement was observed. Spinal bronchogenic cysts were most commonly located at the level of the cervicothoracic spine (15/20, 75.0%). Only 2 cases with spinal bronchogenic cyst in the craniovertebral junction/foramen magnum region were reported. Additionally, no syringomyelia has been described.
previously. During a mean follow-up period of 6.9 months, no recurrence of spinal bronchogenic cyst was noted. The clinical and radiological profiles of previously reported cases were summarized in Table 1.[1–16]

4. Discussion
Bronchogenic cyst is a congenital developmental deformity consisting of approximately 0.7 to 1.3% of all spinal intramedullary tumors. This entity represents a subtype of neurenteric cysts covered with respiratory tract epithelium.[6] The definitive pathogenesis of bronchogenic cysts is currently unclear, whereas 3 theories have been proposed. The 1st hypothesis postulated by Rhaney et al proposed that the bronchogenic cysts are originated from ectoderm, which has potential to differentiate into both endoderm and paraxial mesoderm.[17] The 2nd hypothesis postulated by Bentley et al concluded that the maldevelopment of notochord may result in a fistula between the yolk sac and the amniotic cavity, and then a cyst develops; this hypothesis can explain the ectopic bronchogenic cyst.[18] The 3rd hypothesis claimed by Fallon and Mcletchie assumed that the incomplete separation between the endoderm and ectoderm during differentiation leads to the occurrence of cysts.[19,20] Additionally, Takci et al speculated that congenital tethered spinal cord syndrome might also contribute to the formation of bronchogenic cysts.[21]

The clinical manifestations of spinal bronchogenic cysts are non-specific, which present localization-related mass effect. The most common symptoms include neck and/or back pain, and sensorimotor deficiencies in extremities. In the present case, the patient presented with neck pain and numbness in the upper limbs.

Radiologically, spinal bronchogenic cysts lack typical characteristics, however some signal clues may be suggestive of the diagnosis. Liu et al concluded that spinal bronchogenic cysts are usually hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging.[6] Furthermore, some scholars found the density of spinal bronchogenic cysts on computed tomography (CT) and intensity on MRI may be variable, which are associated with the protein concentration of the cystic contents.[22,23] Our literature review indicates that spinal bronchogenic cysts predominately manifest as hypo- to isointensity on T1-weighted imaging and homogeneous or heterogeneous hyperintensity on T2-weighted imaging. After injection of contrasted medium, no enhancement or only slight enhancement of the cystic wall can be observed.

The differential diagnoses of spinal bronchogenic cysts should include spinal arachnoid cysts, spinal epidermoid cysts, and spinal cystic teratomas. Spinal arachnoid cysts usually show similar signal intensities with cerebrospinal fluid;[24] epidermoid cysts are most commonly found in the lumbosacral segments, and they can present hyperintensity on T1-weighted imaging.[25] However, in a number of cases, radiological identification of these cystic lesions may be challenging, and the definitive diagnosis should depend on histopathological evidence.

The typical pathological features of spinal bronchogenic cysts are pseudostratified ciliated columnar epithelium in the inner wall of the cysts, and in some cases cartilage and smooth muscle can be visible.[26] The reported cases as well as our present case all harbored these characteristics. Immunohistochemical staining can facilitate the diagnosis, which was positive to epithelial membrane antigen (EMA) but negative for glial fibrillary acidic protein (GFAP).[27]

Due to the extremely low morbidity of spinal bronchogenic cysts, the treatment and prognosis have not yet been outlined. These cysts were generally considered to be benign entities, and surgical resection remains the mainstream treatment. Some authors recommended a maximal safe resection; in some cases, the cystic wall may be tightly attached to the spinal cord, and thus
# Table 1

**Literature review of spinal malignant mesothelioma.**

| Author/year | Age/gender | Location | Symptoms | Duration | T1WI | T2WI | Gd-DTPA | Surgical resection | Follow-up period | Recurrence |
|-------------|------------|----------|----------|----------|------|------|---------|-------------------|------------------|------------|
| Ma et al.[1]/2017 | 23 years/F | C4-C7 | Pain in the right upper limb | 1 month | Hypointensity | Hypointensity | No enhancement | STR | 6 months | No |
| | 37 years/F | C3-C6 | Neck pain and numbness in both upper limbs | 2 weeks | Hypointensity | Hypointensity | No enhancement | STR | — | — |
| | 66 years/M | L1-L2 | Lower back pain | — | — | — | Slight enhancement | STR | — | — |
| Vinod et al.[2]/2015 | 45 years/M | L1-T12 | Back pain, weakness and numbness in the lower limbs | 2 months | Hypointensity | Hypointensity | Slight enhancement | GTR | 3 months | No |
| | 37 years/F | C3-C6 | Neck pain and numbness in both upper limbs | 2 weeks | Hypointensity | — | — | STR | — | No |
| | 66 years/M | L1-L2 | Lower back pain | — | — | — | Slight enhancement | STR | — | — |
| Chen et al.[3]/2016 | 24 years/M | L4-L5 | Back pain | 1 month | Isointensity | Hypointensity | No enhancement | STR | — | No |
| | 29 years/M | T9-T10 | Back pain and numbness in the lower extremities | 1 month | Hypointensity | Hypointensity | No enhancement | STR | — | No |
| | 34 years/M | Craniovertical junction | Neck pain and left leg numbness | 6 months | Hypointensity | Hypointensity | No enhancement | STR | — | No |
| | 44 years/F | L4 | Low back pain and weakness in lower extremities | 9 years | Isointensity | Hypointensity | — | GTR | 6 months | No |
| | 55 years/M | T5-T6 | Weakness and numbness in both lower limbs | — | Hypointensity | Hypointensity | — | STR | 12 months | No |
| | 50 years/F | Craniovertical junction | Intermittent occipital headaches, neck pain, syncope attacks, and sensory disturbances in the extremities | — | Hypointensity | Hypointensity | No enhancement | GTR | 3 months | No |
| | 20 years/M | T4 | Back pain, urinary incontinence, numbness in the lower extremities, and increased difficulty in walking. | 6 months | — | Hypointensity | — | GTR | 12 months | No |
| | 17 years/M | T12 | Back pain and paresthesia in both legs | — | — | — | Hypointensity | STR | 6 months | No |
| | 5 months/F | S2 | Skin dimple in the sacral area | — | — | — | Hypointensity | STR | — | — |
| | 28 years/M | L1 | Chronic lumbar, weakness and numbness in both lower limbs | 1 year | — | — | Hypointensity | STR | — | — |
| | 41 years/F | T12-L1 | Chronic lumbosacral pain, sharp pain in the left leg | — | — | — | Hypointensity | STR | 3 months | No |
| | 18 years/M | C2-C3 | Radiating pain and weakness of the right upper limb | 6 weeks | Hypointensity | Hypointensity | — | GTR | 3 months | No |
| | 16 years/M | C1 | Pain in the posterior upper neck region | — | Isointensity | Hypointensity | — | GTR | 12 months | No |
| | 55 years/F | C3-C4 | Pain and paresthesia in her right arm | 2 weeks | — | — | — | STR | 12 months | No |
| | 21 years/F | C5-T2 | Tingling, numbness and diminishing sensation starting in the right arm and leg and later in the left side | 6 weeks | — | — | — | GTR | — | — |
| | 14 years/F | C6-C7 | — | — | — | — | GTR | 11 months | No |

C = cervical, F = female, Gd-DTPA = gadolinium-diethylene triamine pentaacetic acid, GTR = gross total resection, L = lumbar, M = male, STR = subtotal resection, T = thoracic, T1WI = T1-weighted imaging, T2WI = T2-weighted imaging.
gross total resection may be impossible. In this study, we highlighted the value of intraoperative neurophysiological monitoring, which significantly helps protect the functions of spinal cord. A puncture of cysts via the posterior midline approach can help reduce the intracapsular pressure and prevent the irritation of the subarachnoid space. Fievet et al. found bronchogenic cysts with bronchogenic cysts. Che et al. also proposed that spinal bronchogenic cysts might be associated with staphylococcal aureus infection or spontaneous hemorrhage. In the current case and literature review, 9 (45%) cases received gross total resection and 11 (55%) cases received subtotal resection, and no recurrence was noted during the observation period. Although spinal bronchogenic cyst is a low-grade entity with a benign nature, longer follow-up is necessary to make definitive conclusions regarding the prognosis.

5. Conclusion

Spinal intramedullary bronchogenic cyst in the foramen magnum region accompanying with syringomyelia is an extremely rare entity. Preoperative identification based on radiological findings is challenging, and definitive diagnosis depends on histopathological evidence. Appropriate surgical resection is associated with a favorable outcome.

Author contributions

Fan Chen drafted this manuscript. Sascha Marx and Chaochao Zhang analyzed and interpreted the patient data. Junguo Cao, Ying Yu and Dawei Chen evaluated the histopathological images and prepared the figures. All authors read and approved the final manuscript.

Resources: Sascha Marx, Chaochao Zhang, Junguo Cao, Ying Yu, Dawei Chen.

Writing – original draft: Fan Chen.

References

[1] Ma X, Li W, Chen N, et al. Intraspinal bronchogenic cyst: series of case reports and literature review. J Spinal Cord Med 2017;40:141–6.
[2] Chen J, Lai R, Li Z, et al. Case report series and review of rare intradural extramedullary neoplasms-bronchogenic cysts. Medicine 2015;94: e2039.
[3] Solaroglu I, Algin O, Caylak B, et al. Bronchogenic cyst of the craniovascular junction: a case report. Turk Neurosurg 2014;24:284–7.
[4] Vinod K, Nair RP, Deshpajua CK. Bronchogenic intraspinal cyst - a rare case of spinal cystic space occupying lesion. Neurol India 2016;64:1083.
[5] Zou MX, Hu JR, Kang YJ, et al. Bronchogenic cyst of the conus medullaris with spinal cord tethering: a case report and review of the literature. Int J Clin Exp Pathol 2015;8:3997–42.
[6] Liu QP, Zhang JN, Zhang L, et al. An acute case of paraplegia and spinal bronchogenic cyst. J Orthop Sci 2015;20:923–6.
[7] Arnold FM, Neff LL, Anderson KK, et al. Thoracic myelopathy secondary to intradural extramedullary bronchogenic cyst. J Spinal Cord Med 2009;32:595–7.
[8] Yilmaz C, Gulsen S, Sonmez E, et al. Intramedullary bronchogenic cyst of the conus medullaris. J Neurosurg Spine 2009;11:477–9.
[9] Ko KS, Jeun SS, Lee YS, et al. Sacral intraspinal bronchogenic cyst: a case report. J Korean Med Sci 2008;23:895–7.
[10] Chongyi S, Meng Y, Dejun Y, et al. Lumbar intradural extramedullary bronchogenic cyst. Eur Surg Res 2008;40:26.
[11] Baumann CR, Koni D, Glanzel M, et al. Thoracolumbar intradural bronchogenic cyst. Acta Neurochir 2005;147:317–9.
[12] Rato GP, Bhaskar G, Reddy PK. Cervical intradural extramedullary bronchogenic cyst. Neurol India 1999;47:79.
[13] Baba H, Okumura Y, Ando M, et al. A high cervical intradural extramedullary bronchogenic cyst. Case report. Paraplegia 1995;33:228.
[14] Wilkinson N, Reid H, Hughes D. Intradural bronchogenic cysts. J Clin Pathol 1992;45:1032–3.
[15] Ho KI, Tiel R. Intraspinmonary bronchogenic cyst: ultrastuctural study of the lining epithelium. Acta Neuropathol 1989;78:513–20.
[16] Yamashita J, Maloney AF, Harris P. Intradural bronchogenic cyst. Case report. J Neurosurg 1973;39:240–5.
[17] Rhaney K, Barclay GP. Enterogenous cysts and congenital diverticula of the alimentary canal with abnormalities of the vertebral column and spinal cord. J Pathol Bacteriol 1959;77:457–71.
[18] Bentley JF, Smith JR. Developmental posterior enteric remnants and spinal malformations. Arch Dis Child 1960;35:76–86.
[19] Fallon M, Gordon AR, Lendrum AC. Medialstinal cysts of fore-gut origin associated with vertebral abnormalities. Br J Surg 1954;41:520–33.
[20] Mclachngie NG, Purves JK, Saunders RL. The genesis of gastric and certain intestinal diverticula and enterogenous cysts. Surg Gyneco Obst 1954;99:135–41.
[21] Takci E, Sengul G, Keles M. Spinal intramedullary epidymal cyst and tethered cord in an adult. Case report. J Neuroulog Spine 2006;4:506–8.
[22] Cardinale L, Ardissone F, Cataldi A, et al. Bronchogenic cysts in the adult: diagnostic criteria derived from the correct use of standard radiography and computed tomography. Radiol Med 2008;113:385–94.
[23] Meadams HP, Kiricryzk WM, Rosado-De-Christenson ML, et al. Bronchogenic cyst: imaging features with clinical and histopathologic correlation. Radiology 2000;217:441–6.
[24] Wang MY, Levi AD, Green BA. Intradural spinal arachnoid cysts in adults. Surg Neurol 2003;60:49–55.
[25] Karadag D, Karagullue AT, Erden A, et al. MR imaging of a ruptured intraspinal dermoid tumour with fat droplets in the central spinal canal. Aust Radiol 2002;46:444.
[26] Che WC, Zang Q, Zha Q, et al. Lipoma-like bronchogenic cyst in the right chest sidewall: a case report and literature review. Ann Thorac Cardiovasc Surg 2016;22:370.
[27] Savage JJ, Casey JN, McNeill IT, et al. Neuroenteric cysts of the spine. J Craniovertebr Junction Spine 2010;1:58.
[28] Fievet L, D’Journo XB, Gouj JM, et al. Bronchogenic cyst: best time for surgery? Ann Thorac Surg 2012;94:1695–9.
[29] Kirmani B, Kirmani B, Sagolani F. Should asymptomatic bronchogenic cysts in adults be treated conservatively or with surgery? Interact Cardiovasc Thorac Surg 2010;11:649–59.