Clinical Case Report

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Intramedullary bronchogenic cyst in the foramen magnum region accompanied with syringomyelia

A case report and literature review

Fan Chen, MD[a,b], Sascha Marx, MD[b], Chaochao Zhang, MD[a], Junguo Cao, MD[a], Ying Yu, MD[a], Dawei Chen, MD[a,*]

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.

Editor: N/A.

The patient has provided informed consent for publication of the case.

The authors have no funding and conflicts of interest to disclose.

[a] Department of Neurosurgery, First Hospital of Jilin University, Changchun, China, [b] Department of Neurosurgery, University Medicine Greifswald, Greifswald, Germany.

[*] Correspondence: Dawei Chen, Department of Neurosurgery, First Hospital of Jilin University, 71 Xinmin Avenue, Changchun 130021, China (e-mail: professorchen@foxmail.com).

Medicine (2019) 98:5(e14353)

Received: 27 September 2018 / Received in final form: 20 December 2018 / Accepted: 1 January 2019

http://dx.doi.org/10.1097/MD.0000000000014353

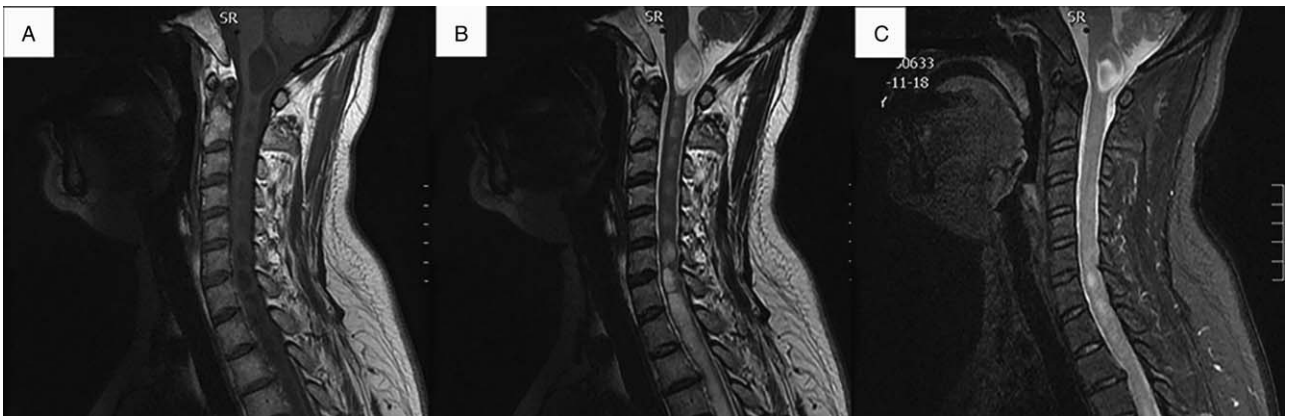

Figure 1. Preoperative magnetic resonance imaging (MRI). (A) Sagittal magnetic resonance T1-weighted imaging showed a cystic lesion with hypointensity in the foramen magnum region. (B) Sagittal T2-weighted imaging showed the lesion was heterogeneously hyperintense. (C) Sagittal contrasted T1-weighted imaging demonstrated heterogeneous enhancement in the cystic wall.

The lesion was hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging; after gadolinium-diethylene triamine 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 craniocervical junction/foramen magnum region were reported.[2,3] Additionally, no syringomyelia has been described

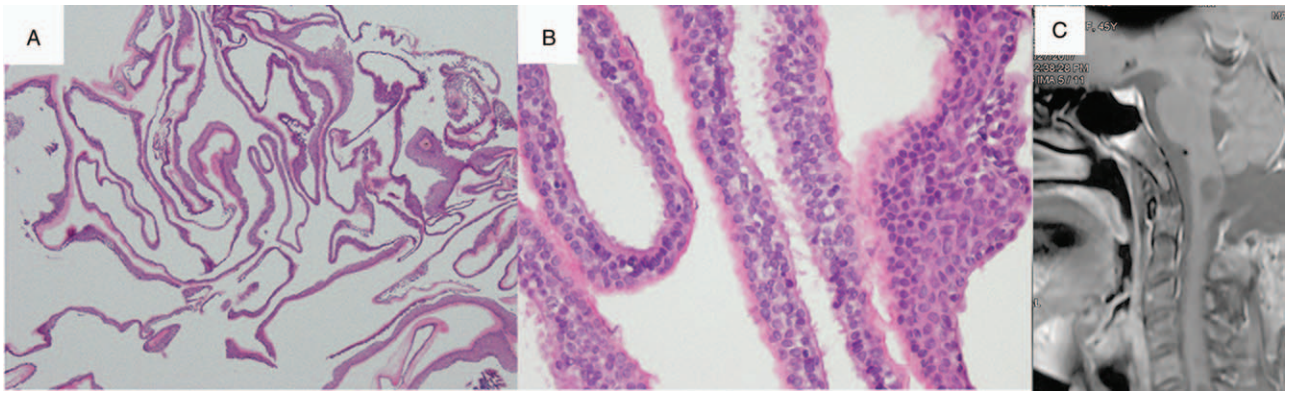

Figure 2. Histopathological findings. Hemotoxylin and eosin (H&E) staining of the resected specimen showed pseudostratified ciliated columnar epithelium with abundant cilia, and there was no cartilage or fibrovascular proliferation in the peripheral interstitium (A, magnification ×40; B, magnification ×400).

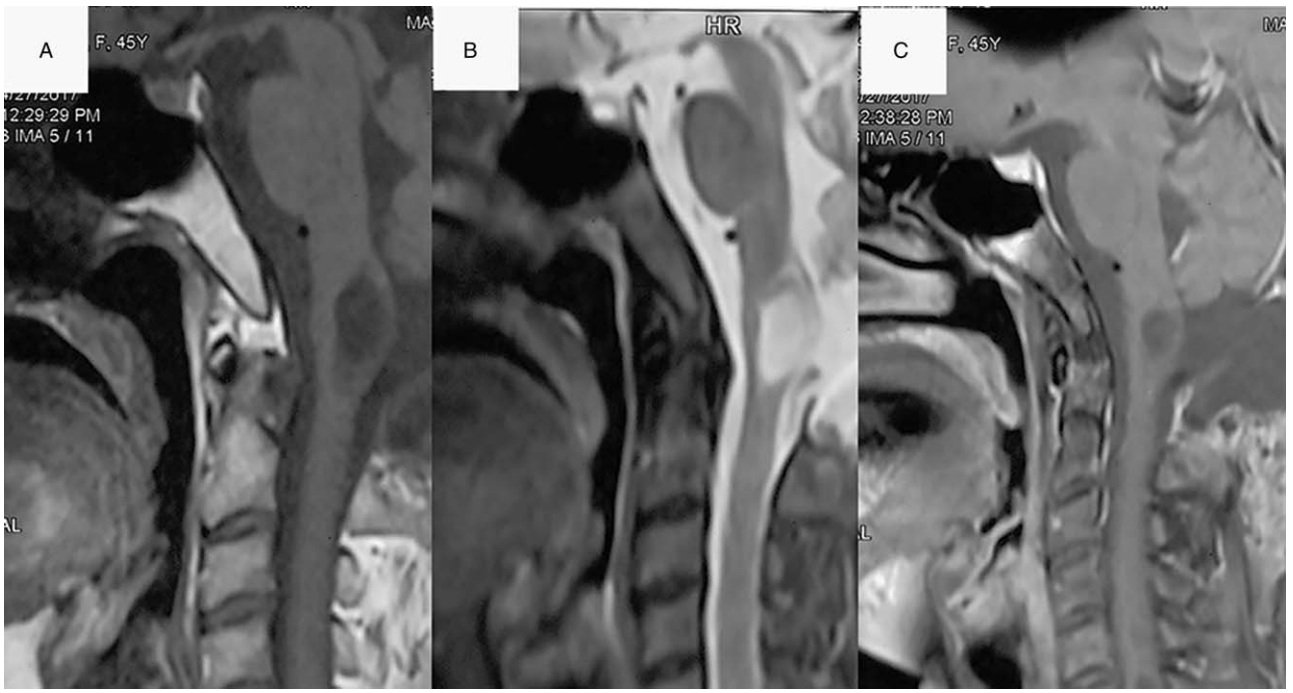

Figure 3. Magnetic resonance imaging (MRI) 3 months after operation. Sagittal magnetic resonance T1-weighted (A), T2-weighted (B), and contrasted T1-weighted imaging showed no recurrence and the syringomyelia disappeared.

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.

					MRI characteristics					
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	Hyperintensity	No enhancement	STR	6 months	No
	37 years/F	C3-C6	Neck pain and numbness in both upper limbs	2 weeks	Hypointensity	Hyperintensity	No enhancement	STR	—	No
	66 years/M	L1-L2	Lower back pain	—	—	—	Slight enhancement	STR	—	No
Vinod et al[4]/2016	45 years/M	T11-T12	Back pain, weakness and numbness in the lower limbs	2 months	—	Hyperintensity	Slight enhancement	GTR	3 months	Yes
Chen et al[2]/2015	24 years/M	L4-L5	Back pain	1 month	Isointensity	Hyperintensity	No enhancement	STR	—	No
	29 years/M	T9-T10	Back pain and numbness in the lower extremities	1 month	Hypointensity	Hyperintensity	No enhancement	STR	—	No
	34 years/M	Craniocervical junction	Neck pain and left leg numbness	6 months	Hypointensity	Hyperintensity	No enhancement	GTR	—	No
Zou et al[5]/2014	44 years/F	L4	Low back pain and weakness in lower extrimities	9 years	Isointensity	Hyperintensity	—	GTR	6 months	No
Liu et al.[6]/2015	55 years/M	T5-T6	Weakness and numbness in both lower limbs	—	Hypointensity	Hyperintensity	—	STR	12 months	No
Solaroglu et al[3]/2014	50 years/F	Craniocervical junction	Intermittent occipital headaches, neck pain, syncope attacks, and sensory disturbances in the extremities	—	Hypointensity	Hyperintensity	No enhancement	GTR	3 months	No
Arnold et al[7]/2009	20 years/M	T4	Back pain, urinary incontinence, numbness in the lower extremities, and increased difficulty in walking.	6 months	—	Hyperintensity	—	GTR	12 months	No
Yilmaz et al[8]/2009	17 years/M	T12	Back pain and paresthesia in both legs	—	—	Hyperintensity	—	STR	6 months	No
Ko et al[9]/2008	5 months/F	S2	Skin dimple in the sacral area	—	Hypointensity	Hyperintensity	—	GTR	9 days	No
Chongyi et al[10]/2008	28 years/M	L1	Chronic lumbago, weakness and numbness in both lower limbs	1 year	—	Hyperintensity	—	STR	—	—
Baumann et al[11]/2005	41 years/—	T12-L1	Chronic lumbovertebral pain, sharp pain in the left leg	—	—	Hyperintensity	—	STR	3 months	No
Rao et al[12]/1999	18 years/M	C2-C3	Radiating pain and weakness of the right upper limb	6 weeks	Hypointensity	Hyperintensity	—	—	3 months	No
Baba et al[13]/1995	16 years/M	C1	Pain in the posterior upper neck region	—	Isointensity	Hyperintensity	—	GTR	12 months	No
Wilkinson et al[14]/1992	55 years/F	C3-C4	Pain and paraesthesia in her right arm	2 weeks	—	—	—	STR	12 months	No
Ho et al[15]/1989	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	—	—
Yamashita et al[16]/1973	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.[9] Herein, 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 liquid to the spinal cord as well. In the present case, the spinal bronchogenic cyst was intramedullary and tightly attached to the spinal cord, and only part of the cystic wall was removed; additionally, we left a fistula between the cyst and the subarachnoid space. Fievet et al found bronchogenic cysts occurring in mediastinum were associated with the risk of transformation to adenoma or rhabdomyosarcoma.[28] Kirmani et al reported a malignant transformation rate of 0.7% in adults with bronchogenic cysts.[29] Che et al also proposed that mediastinal bronchogenic cysts might have a risk of malignant transformation or spontaneous infection.[26] Chen et al reported spinal bronchogenic cysts might be associated with staphylococcus aureus infection or spontaneous hemorrhage.[2] 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.

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