Clinical Case Report

Medicine®

OPEN

An intradural extramedullary bronchogenic cyst in the thoracolumbar spine

A case report

Hyeok Dong Lee, MD[a], Seung Hoon Han, MD, PhD[a], Si-bog Park, MD, PhD[a], Yong Ko, MD, PhD[b], Kyu Hoon Lee, MD, PhD[a,*]

Abstract

Rationale: We report the symptoms beginning with weakness and the clinical courses of a patient who was diagnosed with an intradural extramedullary bronchogenic cyst.

Patient concerns: The patient was a 44-year-old man visited the Department of Physical Medicine and Rehabilitation for walking difficulties characterized by limping due to muscle weakness of left lower extremity for 5 months and atrophy in left calf muscle.

Diagnoses: Lumbar spine MRI was repeated, since radiating pain in the left hip and posterior thigh with low back pain developed 16 months later. Intraspinal mass of T12 and L1 levels that was not found in the first MRI was newly found in the follow-up MRI.

Interventions: Total tumor removal was conducted with laminectomy. It was finally diagnosed as an intradural extramedullary bronchogenic cyst on the basis of the pathological analysis results.

Outcomes: His left calf circumference was increased compared to before surgery the radiating pain also disappeared.

Lessons: If the patient's MRI findings are not correlated with the electrophysiologic and physical examination findings, additional MRI should be accompanied with other tests for an early detection.

Abbreviations: EMG = electromyography, MRI = magnetic resonance imaging.

Keywords: bronchogenic cyst, pain, radiculopathy, weakness

1. Introduction

A bronchogenic cyst is a congenital malformation derived from the primitive foregut. It is an endodermal cyst surrounded by respiratory tract epithelium and is mainly found in the sternum, skin, stomach, and pericardium.[1] However, bronchogenic cysts in the spinal canal are rare.[2,3] In fact, few cases of intradural extramedullary bronchogenic cyst were reported globally. In most cases, pain was the chief complaint, and none of the patients complained of weakness and atrophy. In the thoracolumbar area, far fewer cases of bronchogenic cyst were reported.[4] Baumann et al have reported an bronchogenic cysts in the thoracolumbar area for the first time; however, intradural extramedullary mass in the T12 and L1 levels was found via magnetic resonance imaging (MRI) performed after left leg pain suddenly occurred in a patient with a known spina bifida occulta at 2 lumbar levels (L4/L5) with chronic nonradiating lumbovertebral pain.[4] Conversely, in the case of the present study, atrophy and weakness of left lower extremity were chief complaints. In the follow-up outpatient clinic visits, the calf circumference diameter was measured, and electrophysiologic examination was conducted twice. Furthermore, unlike previously reported cases, there was no abnormal finding on the MRI performed at the first visit, and the symptom worsened instead of recovering; 16 months later, an intradural extramedullary mass was found on the MRI. Herein, we report the symptoms beginning with weakness, the result of the examinations, and the clinical courses of a patient who was diagnosed with an intradural extramedullary bronchogenic cyst, which is a rare intraspinal mass in thoracolumbar area, based on 16 months of hospital visit, treatment, and clinical course.

2. Case presentation

The patient was a 44-year-old man who had been taking medications for asthma for a year, but has no specific medical history, except that he visited the Department of Physical Medicine and Rehabilitation for walking difficulties characterized

Editor: N/A.

Written informed consent was obtained for publication of this case report from the patient.

The Ethics Approval/Institutional Review Board (IRB) is not needed because this is not a case of new intervention is performed.

The authors alone are responsible for the content and writing of the article.

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

[a] Department of Rehabilitation Medicine, [b] Department of Neurosurgery, Hanyang University College of Medicine, Seoul, Republic of Korea.

[*] Correspondence: Kyu Hoon Lee, Department of Rehabilitation Medicine, Hanyang University Hospital, 222-1 Wangsimni-ro, Seongdong-gu, Seoul 04763, Republic of Korea (e-mail: dumitru1@hanyang.ac.kr).

Medicine (2017) 96:50(e9263)

Received: 12 November 2017 / Accepted: 22 November 2017

http://dx.doi.org/10.1097/MD.0000000000009263

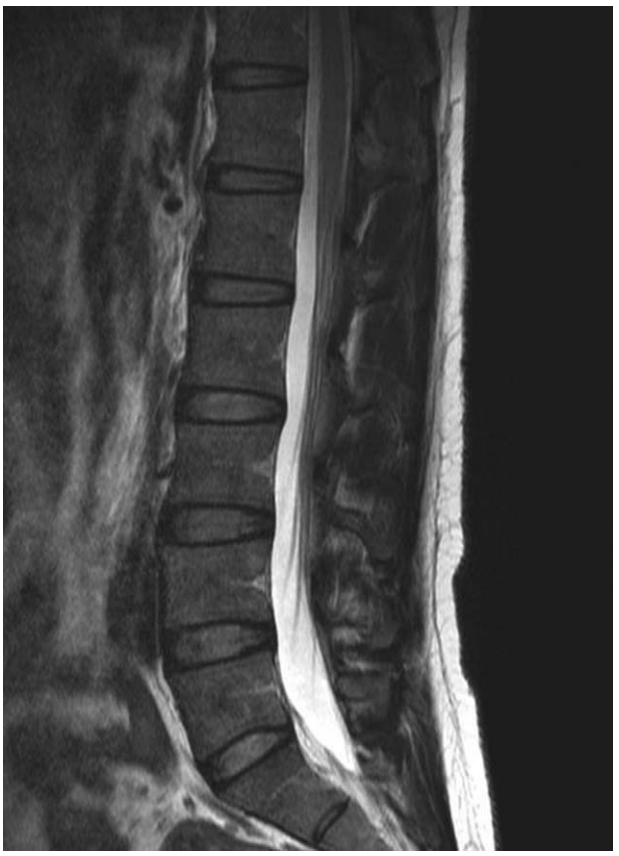

Figure 1. Sagittal T2-weighted magnetic resonance image showing mild disc protrusions.

by limping due to muscle weakness of left lower extremity for 5 months and atrophy in left calf muscle. At that time, his right and left calf circumference was 39.5 and 35.5 cm. The muscle strength of the left hip flexor, ankle dorsi flexor, and ankle plantar flexor assessed via manual muscle testing was reduced as good grade, whereas the patient's bladder and bowel function was normal. Two weeks later, electromyography (EMG) and lumbar spine MRI were performed. According to the result of the EMG, abnormal spontaneous activities were found in the left gastrocnemius, abductor digiti minimi, and abductor hallucis muscle. The diagnosis was left lumbosacral radiculopathy (mainly S1). However, there was no specific abnormal finding in the lumbar spine MRI, except for L1–2, L2–3, and L4–5 disc mild protrusions (Fig. 1). Strengthening exercises and exercise education were conducted. He visited the hospital and was evaluated 4 times in a year, as shown in Table 1. The calf circumference measured a year later improved because of regular gait training and strengthening exercises; however, his left calf circumference was still reduced (right, 42.5 cm; left, 36.5 cm). Moreover, abnormal spontaneous activities in the needle EMG were shown in the left gastrocnemius, peroneus longus, abductor digiti minimi, and abductor hallucis brevis muscles, and neuropathic patterns (increased amplitude, polyphasic, and reduced recruitment patterns) were found in the motor unit action potential analysis, according to the result of the follow-up EMG. Left lumbosacral radiculopathy (mainly S1, chronic state) was considered on the basis of the examination findings.

Lumbar spine MRI was repeated because radiating pain in the left hip and posterior thigh with low back pain developed 16 months later. Hypointensity on T1WI in the T12 and L1 levels and spinal cord tumor with hyperintensity on T2WI were found on the MRI (Fig. 2). Therefore, the patient was admitted to the neurosurgery department in our hospital and underwent excision of the spinal cord mass. Total tumor removal was conducted with laminectomy of the T12 and L1 levels, and an ~3-cm-sized yellowish mass was excised. The mass was ovoid in shape and had a cystic component; it was sent to the pathology department for biopsy. An endodermal cyst with a respiratory epithelium (cytokeratin 7—positive, p63—positive, and TTF-1—positive) was confirmed on the basis of the pathological analysis results and was finally diagnosed as an intradural extramedullary bronchogenic cyst (Fig. 3). On the MRI performed a month after surgery, the tumor was not found. His left calf circumference was still reduced compared with his right calf circumference, but was higher than that prior to the surgery; the tingling sensations also disappeared (Table 1).

3. Discussion

Histologically, congenital intraspinal cysts can be classified into 3 types: epithelial, mesenchymal, and mixed types. Epithelial cysts can be further divided into endodermal (neurenteric and

Table 1

Summary of evaluations and managements during the outpatient clinic follow-ups.

Date	Evaluations	Managements
2014.08.28	Calf circumference (39.5/35.5)	Gait training
		Strengthening exercise
2014.09.11	Calf circumference (39.5/34.5)	Gait training
	MRI: Lumbar disc protrusions	Strengthening exercise
	NCV and EMG: Suggestive of left lumbosacral radiculopathy (mainly S1)	Exercise education
2014.10.23	Calf circumference (40.0/36.0)	Iliolumbar ligament injection
2015.01.31	Calf circumference (39.5/35.0)	Gait training
		Strengthening exercise
2015.04.11	Calf circumference (43.5/37.5)	Gait training
		Strengthening exercise
2015.08.10	Calf circumference (42.5/36.5)	Gait training
	NCV and EMG: Suggestive of left lumbosacral radiculopathy (mainly S1, chronic state)	Strengthening exercise
2015.12.24	Calf circumference (40.0/33.0)	Facet joint block (both L4-L5 and L5-S1 joints)
2015.12.29	MRI: Thoracolumbar intraspinal mass	Laminectomy T12, L1 and total tumor removal
2016.04.09	Calf circumference (42.0/33.6)	

EMG = electromyography, MRI = magnetic resonance imaging, NCV = nerve conduction velocity.

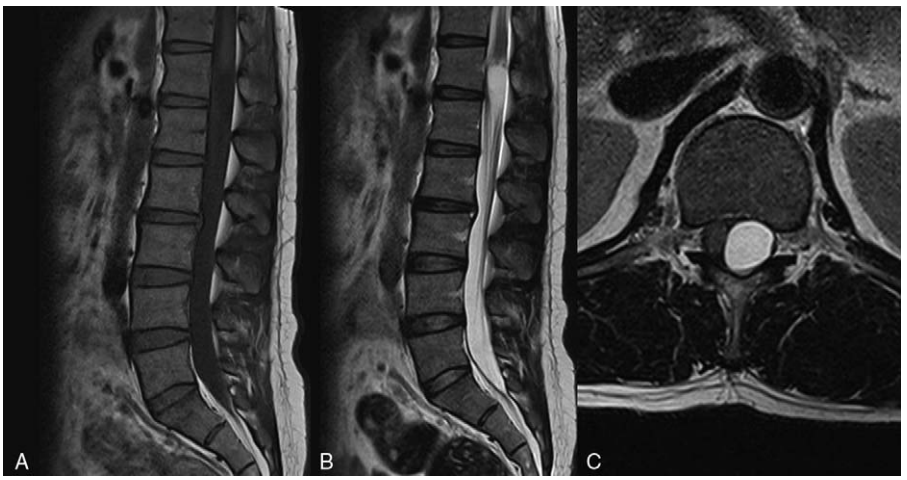

Figure 2. Magnetic resonance image of the thoracolumbar spine. (A) A sagittal T1-weighted magnetic resonance image demonstrates a hypointense cystic lesion, and (B) sagittal and (C) corresponding axial T2-weighted magnetic resonance images demonstrate hyperintense cystic lesions at the T12 to the L1 level.

enterogenous) and ependymal cysts.[4] Endodermal cysts, which are uncommon lesions, account for 0.5% of spinal cord space-occupying lesions.[5] There were previous various views on the definition of endodermal cysts; however, Brun and Saldeen defined the characteristics of typical endodermal cysts as follows: has a gastrointestinal and/or respiratory epithelium, and mainly occurs in the cervical and thoracic regions.[6] Therefore, the term bronchogenic cyst indicates tissues surrounded by the respiratory tract epithelium around endodermal cysts. The exact mechanism has not been found yet. Generally, it is considered that bronchogenic cysts are caused by anomalous embryological connections between the primitive foregut and developing neural tube and are related to splitting or reduplication of intervening notochords.[7] According to existing reports on bronchogenic cysts, these cysts were diagnosed for the first time in the first and second decades of life (61.5%) and often found in men (58.3%)[8] in most cases. The most common symptom was a local mass effect by the tumor and characterized mostly by radiating pain.

Bronchogenic cysts are very rare in the intraspinal area. Most of them are found in the intradural extramedullary area. There are few differences between the previous case of thoracolumbar intradural extramedullary bronchogenic cyst reported by Baumann et al. and our case. In a previous case, an intraspinal tumor was found via MRI immediately after neurological examinations and physical evaluations after radiating pain on the left lower extremity occurred. Laminectomy and cystic mass removal were conducted after it was diagnosed.

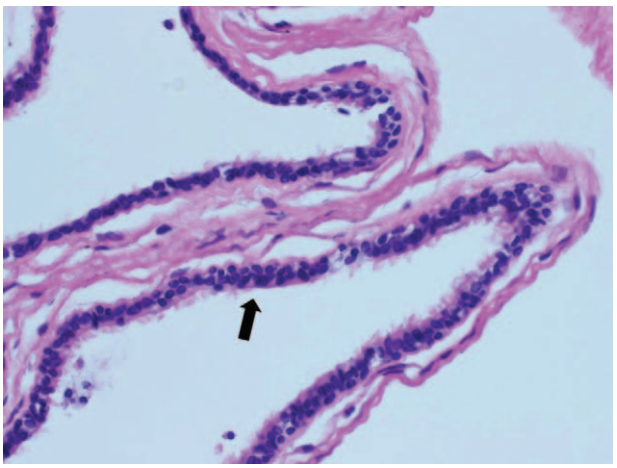

Figure 3. Histologically, a cyst lined with a respiratory-type pseudostratified columnar ciliated epithelium (arrow) is a characteristic of a bronchogenic cyst (hematoxylin and eosin, ×200).

Conversely, the first symptoms of the case in this study were calf atrophy and motor weakness, which were different from those of other cases. Pain developed after 16 months in this case. The patient underwent serial checkups for 16 months at the outpatient clinic, and the progress of his condition was measured through calf circumference and muscle strength assessments. Moreover, Bauman et al's case showed developmental anomalies, whereas our case showed no anomalies. Therefore, it was difficult to predict congenital malformations. None of the previous studies conducted EMG; however, radiculopathy in our case was found via EMG. In addition, there was no special abnormality, such as tumor mass on the MRI conducted at the first outpatient clinic visit, except for disc protrusion. Generally, these findings may lead to the exclusion of other diagnoses because no abnormality was shown on the first MRI. However, as the patient newly felt radiating pain on his pelvic limb rather than recover from the symptom, we repeated MRI; as a result, the intraspinal mass was found. If new symptoms had not occurred, follow-up MRI would not have been performed in our case. Furthermore, it is difficult to consider other diagnoses if no changes are seen in the symptoms. Therefore, performing follow-up MRI in addition to other tests, such as physical examination and EMG, when there is no improvement in the symptoms for a long period despite no abnormality on the first MRI is needed. Furthermore, additional MRI could be helpful when the first MRI findings do not correlate with other test findings.

Intradural extramedullary mass is mainly found via MRI. MRI is regarded as the most suitable tool to examine spinal cord tumors owing to its advantages of improved resolution, fewer artifacts, and multiplanar images. Based on a previous research, bronchogenic cysts on MRI have isointense or hypointense signals on T1W1 and hyperintense signals on T2W1; furthermore, in many cases, the cysts are difficult to be enhanced using intravenous contrast injection. Computed tomography (CT) is necessary to diagnose other congenital bony anomalies.[8] Total surgical resection is required to treat bronchogenic cysts. There is

a high risk of recurrence in partial resection owing to the possibility of remnant tissue differentiations.[9] There is no reported recurrence in gross total resection; however, partial resection showed a 63% rate of recurrence.[10] Most of the patients' surgical outcomes were good, and cystic mass was not found on the follow-up MRIs. In conclusion, spinal bronchogenic cysts are very rare and even rarer in the thoracolumbar or lumbar spinal area than in the cervical spinal area. In other cases, pain was the symptom; however, motor weakness and muscle atrophy were shown without pain only in our case. When lumbar disc protrusions and lumbosacral radiculopathy were found via MRI and EMG, respectively, there is a high possibility that such a condition would be diagnosed as herniation of the lumbar disc or spinal stenosis. However, other diagnoses should always be considered. Furthermore, if the symptoms are not sufficiently explained by the findings of the early MRI or do not improve for a long period, follow-up MRI should be considered. Moreover, although the case seems to have similar symptoms with S1 radiculopathy according to the EMG findings, the symptoms can be caused by the mass effect due to the tumor in a higher level. Therefore, the possibility of congenital malformations should be considered as well.

The pathophysiology of intradural extramedullary bronchogenic cysts, including its progression rate, is not clearly defined. Therefore, further studies are needed to clarify the pathophysiology of intradural extramedullary bronchogenic cysts. Besides, if the patient's MRI findings are not correlated with the electrophysiologic and physical examination findings, additional MRI should be accompanied with other tests for an early detection.

References

[1] Ko CL, Lin HJ, Foo NP, et al. Bronchogenic cyst. J Emerg Med 2010;39:650–1.
[2] Chongyi S, Meng Y, Dejun Y, et al. Lumbar intradural extramedullary bronchogenic cyst. Eur Surg Res 2008;40:26–8.
[3] Rao GP, Bhaskar G, Reddy PK. Cervical intradural extramedullary bronchogenic cyst. Neurol India 1999;47:79–81.
[4] Baumann CR, Konu D, Glatzel M, et al. Thoracolumbar intradural extramedullary bronchiogenic cyst. Acta Neurochir 2005;147:317–9.
[5] Sun JJ. Classification, mechanism and surgical treatments for spinal canal cysts. Chin Neurosug J 2016;2:7–17.
[6] Brun A, Saldeen T. Intraspinal enterogenous cyst. Acta Pathol Microbiol Scand 1968;73:191–4.
[7] Kulkarni V, Daniel RT, Haran RP. Extradural endodermal cyst of posterior fossa: case report, review of the literature, and embryogenesis. Neurosurg 2000;47:764–7.
[8] Chen J, Lai R, Li Z, et al. Case report series and review of rare intradural extramedullary neoplasms—bronchiogenic cysts. Medicine (Baltimore) 2015;94:2039–43.
[9] Gauden AJ, Khurana VG, Tsui AE, et al. Intracranial neuroenteric cysts: a concise review including an illustrative patient. J Clin Neurosci 2012;19:352–9.
[10] Garg N, Sampath S, Yasha TC, et al. Is total excision of spinal neurenteric cysts possible. Br J Neurosurg 2008;22:241–51.