Review

Intraspinal bronchogenic cyst: Series of case reports and literature review

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Purpose: Spinal bronchogenic cysts (SBCs) are rare congenital lesions. The clinical and imaging characteristics and treatment of SBCs are not well known. We studied three cases of SBCs retrospectively, which were registered in our department and analyzed eight case reports which were all published in English, focusing on providing a deeper knowledge of SBCs.

Methods: Three patients with SBCs registered in our department were retrospectively reviewed. Eight reported SBCs cases published from 1992 to 2015 were enrolled in our study. Imaging diagnosis was confirmed by computed tomography (CT), MRI, and computed tomography angiography (CTA). All of our patients and reviewed cases had undergone surgical resection and the final diagnosis was made by pathological examination.

Results: Five lesions were located at the cervical spinal canal. Most patients presented with pain in the limbs and back, which might be related to compression of the spinal cord and the reduced blood supply of the anterior spinal artery. The signal intensity on MRI was correlated with cystic fluid traits to a large extent. Seven lesions were partially removed because of the adhesions to the nearby spinal cord. All reported cases had no recurrence in the later follow-up.

Conclusions: SBCs can occur anywhere in the spinal canal, but they are more likely to present at the cervical canal and might be present along with some developmental malformations of the spine. We emphasize the role of CT and MRI findings in the disease diagnosis. It is recommended that the lesion should be removed as completely as possible.

Keywords: Computed tomography, Fused cervical vertebra, Laminectomy, Magnetic resonance imaging, Spinal bronchogenic cyst

Introduction

Bronchogenic cysts are considered to be a congenital abnormality derived from the developing endoderm, predominantly lined with pseudostratified ciliated columnar epithelium.[1,2] The literatures review about spinal bronchogenic cysts (SBCs) showed that cervical and upper thoracic segments are their predilection sites, and almost all of the cysts have an intradural extramedullary localization. Several case reports of SBCs suggested that the lesions enlarge slowly and lead to pain, numbness, weakness of limbs or back and even paralysis when they compress the nerve roots or spinal cord.[3–8] None of the complications secondary to ruptured cysts were reported, such as chemical meningitis, abscess formation, and hydrocephalus. It is difficult to make the correct diagnosis before surgery due to its extremely low morbidity and non-typical signs, symptoms and imaging findings.

The origin of SBCs is not fully understood. There are various hypotheses about the pathogenesis of SBCs, but the split notochord syndrome theory is widely accepted because it explains the association between SBCs and vertebral anomalies.[3] One of our cases presents fused cervical vertebrae, which strongly supports this hypothesis.

Does the purity of SBC's fluid result in a variety of MRI signal intensities on either T1WI or T2WI or both? What is the best surgical intervention to remove the lesion and relieve the clinical symptoms? We studied three cases of SBCs retrospectively which were confirmed by pathological examination and reviewed the published literatures to find out the imaging characteristics and best management strategy of SBCs.

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Case reports

Case 1

A 23-year-old female presented with pain in the right upper limb for one month. She had a claw deformity of the left hand and talipes equinovarus of the right foot since childhood. The physical examination revealed anisocoria (left pupil was 2 millimeter in diameter while the right pupil was 3 millimeter in diameter), enophthalmos, blepharoptosis and hypohidrosis on the left side face. Muscular tension and strength of limbs were normal. The Magnetic Resonance Imaging (MRI) report from the local hospital revealed a cystic mass in the cervical spinal canal.

The MRI showed an intradural extramedullary cystic lesion located at the C4–C7 level stretching to extraspinal space through the left C5–C6 intervertebral foramen. Some lipomatous tissue signal was observed around the mass. The adjacent spinal cord was visibly compressed. The lesion had no enhancement on the gadolinium-enhanced MRI (Fig. 1A-C). As a result, the imaging diagnosis was considered to be a schwannoma. Spiral-CT three-dimensional reconstruction demonstrated that C4-T1 vertebraes were fused, accompanied by an obvious scoliosis (Fig. 1D).

In order to relieve the compressive symptoms, the patient underwent surgery under general anesthesia. During the operation, it was observed that a mixed solid cystic mass was located at the C5 level, and the spinal cord was compressed towards the right in front of the mass. Colorless and clear cystic fluid was observed after the capsule was opened. The arachnoid membrane hyperplasia on the surface of the mass resulting in adhesion with the spinal cord made it difficult to remove completely. After resecting a part of the intraspinal mass, the spinal cord compression was relieved. But the extraspinal part was not removed in order to avoid further surgical trauma. The pathological results demonstrated that the cystic wall was lined with pseudostratified ciliated columnar epithelium, which supported the diagnosis of a SBC (Fig. 1E).

The symptoms were relieved after removing the intraspinal part of the mass, and the postoperative MR images showed that the extraspinal part of the mass still existed. The lesion has not recurred during a 6 month follow-up.

Case 2

A 37-year-old female with neck pain and numbness in both upper limbs for two weeks was admitted in the hospital. The neurological examination showed hypoesthesia in the posterior part of the neck, shoulders and arms.

MRI showed an intradural extramedullary cystic lesion located at the level of C3–6 in front of the spinal cord. The size was about 5 centimeter. It presented hyperintensity on T2WI and hypointensity on T1WI similar with the cerebrospinal fluid (CSF). The lesion was not enhanced on the gadolinium-enhanced MRI. The final imaging diagnosis was made an arachnoid cyst. Spiral-CT three-dimensional reconstruction revealed that the lesion was located at C2–7 and the CT attenuation value was 21 Hounsfield, with no sign of spinal deformity.

Laminectomy of C3–5 was performed in order to remove the lesion under general anesthesia. The lesion was well exposed by a longitudinal opening of the dura mater and a thin cystic wall was observed. The middle ventral parts of the capsule were adhered to the dorsal cervical cord. Thus, only the superior and inferior parts could be resected, while the adhered parts were residual. There was no complication during and after the operation.

The pathological examination demonstrated a pseudostratified columnar ciliated epithelium lined along with internal layer of the cyst, which supported the diagnosis of a bronchogenic cyst.

The postoperative course of the patient was favorable. The neck pain and numbness of upper limbs were relieved and sensation of arms was recovered. A repeated MRI examination did not show any mass in the cervical spinal canal and the spinal cord was along the midline again.

Case 3

A 66-year-old male had been feeling lower back pain which worsened after 10 days of activity. Related physical examinations didn't show any abnormal signs.

The local hospital's MRI report revealed an intradural extramedullary lesion in the lumbar spinal canal. An enhanced-MRI scan showed a slightly enhanced extramedullary lesion at the L1–2 level and the spinal cord was compressed forward. The final imaging diagnosis was considered as a schwannoma.

A white, fluid-containing, $3\times3\times2cm^3$ cystic lesion was identified at the dorsal part of the spinal cord when the dura mater was longitudinally opened. Gelatinous and viscous liquid was observed when the cyst was punctured. Part of the cystic wall was tightly attached to the dorsal part of the spinal cord, thus the lesion was partially removed. No complications were found during and after the surgery. Re-examination by MRI did not reveal any recurrent signs.

A SBC was confirmed by pathological examination, which showed the cyst wall was lined with pseudostratified columnar ciliated epithelium.

Discussion

WHO defines enterogenous cysts as a kind of cyst which is lined with gastrointestinal, respiratory, stratified

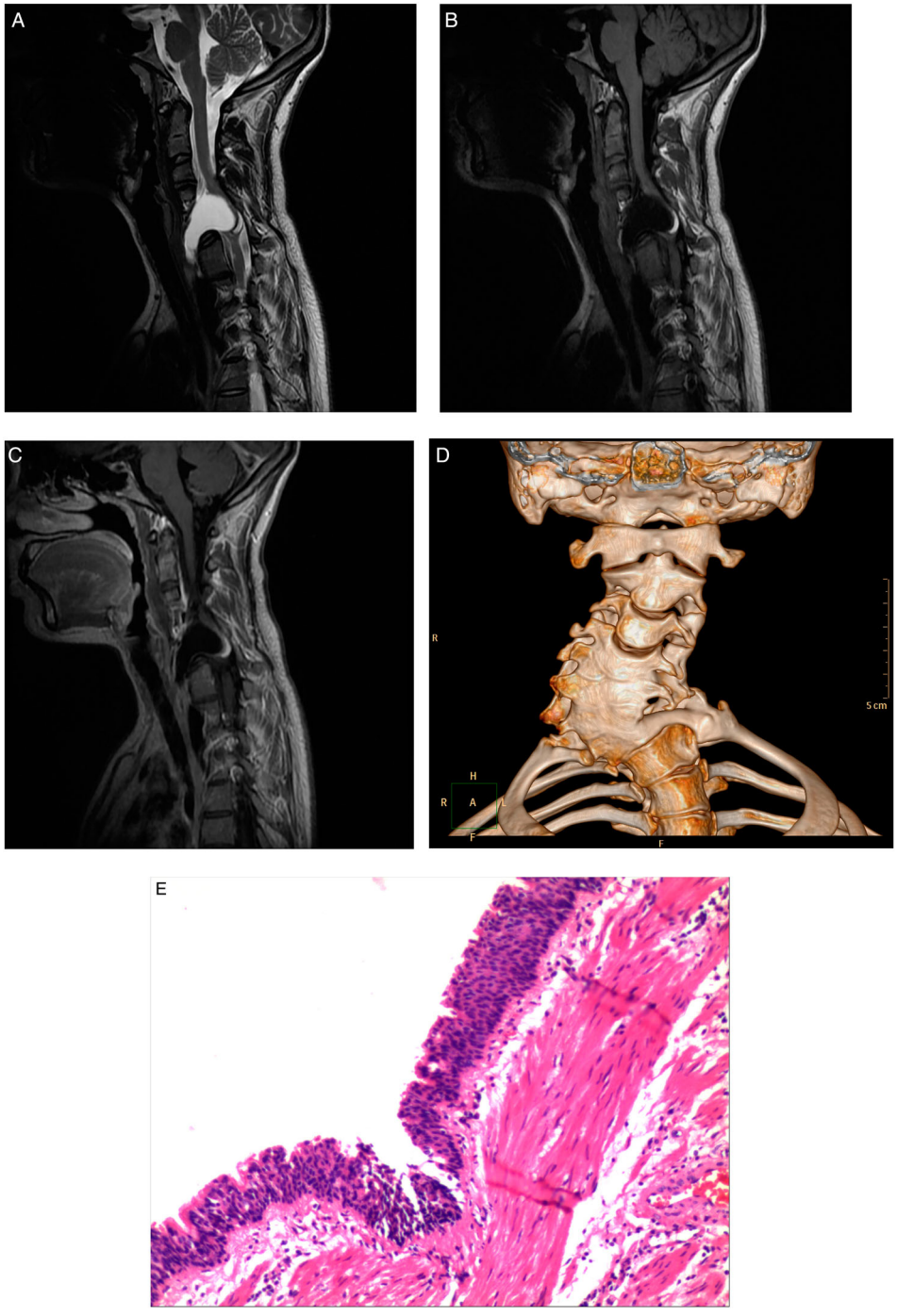

Figure 1 **A Sagittal T2-weighted MRI showing a hyperintense lesion at C4-7 stretched over the spinal canal, which compressed the spinal cord. B The lesion presented with a hypointense and a lunular hyperintense signal on T1-weighted MRI, which was still high on T2WI. C Contrast-T1WI suggested the lesion was not enhanced and the lunular high signal was still present. D The fused C4-7 vertebral bodies were presented well at the spiral-CT three-dimensional reconstruction image. E The pathological image under a microscope (HE 10×) revealed the cyst wall was lined with pseudostratified ciliated columnar epithelium.**

squamous or epithelial cells and secretes mucus protein.[1] If it is lined with respiratory epithelium, the term bronchogenic cyst is used. If the cyst appears in the spinal canal, it is known as spinal bronchogenic cyst (SBC).[2] Thus, the bronchogenic cyst can be considered as a subtype of neurenteric (enterogenous) cyst. Enterogenous cysts represent 0.7–1.3% of total spinal cord tumors.[9,10] The origin of a bronchogenic cyst is not completely known, but it has been proposed to result from three hypotheses.[3] At first, it's assumed that the endoderm and ectoderm do not separate completely during differentiation of the inner cell mass.

Secondly, the cyst is considered to originate from ectoderm because of its potential to form endoderm and paraxial mesoderm. Lastly, it is attributed to the split notochord syndrome which can explain the ectopic bronchogenic cyst. When the duplication or separation of the notochord is incomplete, ectopic cysts are generated and present the characteristics of any part of the foregut.[4]

It is difficult to diagnose spinal bronchogenic cyst before surgery and the final diagnosis depends on the pathological examination. A research confirmed that six cell types were identified, including ciliated cells, nonciliated cells, goblet cells, basal cells, Kulchisky's cells and undifferentiated cells, which were similar to the human bronchial epithelium of the upper respiratory tract.[11] In our case, similar pathological features were found. Beside these typical cells, some fat tissues were also found around the cyst, which were corresponding to the high signal around the cyst on both T1WI and T2WI.

SBCs grow slowly that do not cause any symptoms until they compress the spinal cord. From the eight published studies (Table 1), six cases presented with pain in the limbs and back, three with limb weakness, two with numbness, and only one with paresthesia. A 5-month-old baby was admitted to the hospital and underwent surgery on account of a skin dimple in the sacral area.[12] Wang *et al*. studied 21 adult patients with intradural spinal arachnoid cysts and suggested that ventral cysts presented more commonly with weakness and myelopathy, while dorsal cysts were more inclined to lead to neuropathic pain and numbness.[13] In addition, the symptoms may be explained by local compression of spinal tracts, and ventral cysts may compress the anterior spinal artery to cause weakness and myelopathy.[13]

It is reported that fluid within the bronchogenic cysts contains a mixture of water and proteinaceous mucus.[14,15] With the increasing concentration of protein and mucus, the fluid presents from a thin liquid to a very viscous and mucoid component, which may be related to various densities on CT images and signal intensity on MR images.[15] By reviewing the case reports of SBCs from 1992 to 2015, six cysts presented with homogenous hyperintense on the T2WI and hypointense on T1WI, and only one case showed heterogeneous hyperintense on T2WI and hypointensity on T1WI which could be explained by the surgery findings of heterogeneous cystic fluid. One case had lipomatous tissue adhered to the caudal end of the cystic mass, which was observed on MRI and later was confirmed after surgical and pathological examination (Table 1). One of our cases also found some fat signals near the cyst on MRI. Whether the lipomatous tissue adhering to the cyst is a specific sign of SBCs or not, may need more literature reviews and studies. All of the cases reported that the masses only grew inside the intraspinal canal, while one of the case admitted in our hospital grew across the intervertebral foramen resulting in a misdiagnosis of schwannoma which is more common in the spinal canal and tends to grow along the intervertebral foramen.

Since SBCs are rare and have no specific characteristics, the misdiagnosis rate is high. The following diseases should be differentiated. (i) Spinal arachnoid cysts are more commonly found in the dorsal part of thoracic spinal canal and present with a CSF signal-intensity on all MRI sequences.[13,16] (ii) Spinal dermoid cysts and epidermoid cysts can be intramedullary, intradural extramedullary or extradural and mostly occur at the lumbosacral region. MR images can present with different intensities due to various lipids in the cystic fluid (cholesterol crystals, lipid metabolites and keratin).[17] High signal on DWI is helpful for differentiating from other cystic diseases.[18] (iii) Spinal mature cystic teratomas are more common in children and adolescents and have no special imaging characteristics, thus the final diagnosis depends on the pathological examination.[19]

Because there are no sufficient reports available, the most effective management of SBCs is still poorly understood. Several case reports of mediastinal and retroperitoneal bronchogenic cysts demonstrated the potential of a malignant transformation.[20,21] SBCs are always accompanied by different kinds of spinal deformities, which could be symptomatic and reduce the patient's quality of life. Surgical resection is thought to be the best method of relieving the compressed symptoms.[7] All of the published cases have undergone laminectomy for the purpose of removing the cyst. However, the residual cyst wall might be associated with recurrence.[22] According to a research about neurenteric cysts near the lower clivus, the authors established that the complete excision of the wall by aspiration and ideal marsupialization were closely associated with a favorable prognosis.[23] From the reviewed cases, only in half of the case reports, cysts were removed completely (Table 1). The proportion of total excision of SBCs (50%) was a little higher than the reported excision of spinal neurenteric cysts (36%).[24] A study of the neurenteric cysts presented that the severity of the adhesion with nearby neural structures had a positive correlation with the size of the cyst and the thinness of the wall.[25]

Table 1 Summary of reported cases with SBCs

Authors	Year	Age/Sex	Symptoms	Location	MRI findings	Fluid of cysts	Resection
Liu et al.[8]	2015	55 years/M	Progressive weakness and numbness in both lower limbs	Thoracic intradural extramedullary	Heterogeneous hyperintensity on T2WI and hypointensity on T1WI	Mostly clear and colorless, the rest was brownish yellow and viscous.	Partial resection
Arnold et al.[5]	2009	20 years/M	Back pain and lower-extremity weakness	Thoracic intradural extramedullary	Homogenous hyperintensity on T2WI and hypointensity on T1WI	—	Total resection
Chongyi et al.[7]	2008	28 years/M	Chronic lumbago and progressive weakness and numbness in both lower limbs	Lumbar spinal canal	Homogenous hyperintensity on T2WI	Yellow viscous liquid	Partial resection
Ko et al.[12]	2008	5 months/F	Skin dimple in the sacral area	Sacral intradural extramedullary	Homogenous hyperintensity on T2WI and hypointensity on T1WI	Clear and colorless liquid	Total resection
Baumann et al.[3]	2005	41 years/M	Chronic nonradiating lumbovertebral pain	Thoracolumbar intradural extramedullary	Hyperintensity on T2WI. Lipomatous tissue adherent to the caudal end of the cystic mass	Gelatinous cream-colored liquid	Partial resection
Rao et al.[1]	1999	18 years/M	Radiating pain and progressive weakness of the right upper limb	Cervical intradural extramedullary	Similar to CSF on both T1WI and T2WI	Clear fluid	Total resection
Baba et al.[26]	1995	16 years/M	Intractable neck pain	Cervical intradural extramedullary	Intermediate signal intensity on TIWI and slightly higher on T2WI	Slightly opaque and colorless	Total resection
Wilkinson et al.[6]	1992	55 years/F	Increasing pain and paresthesia in the right arm	Upper cervical spinal canal	—	Viscous, opaque, and white liquid	Partial resection

The prognosis of SBCs is comforting and no recurrence was reported in all of our cases. Outcomes of all the three cases presented no recurrent signs in MRI examination.

Conclusion

SBCs are rare intraspinal extramedullary lesions which can cause symptomatic spinal cord compression. The most common imaging presentation on MRI is homogenous hypointense on TIWI and hyperintense on T2WI and the lipomatous tissue may be a helpful sign for diagnosis. The recommended management is total resection of the cyst although half of the reported lesions can not be completely removed.

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Conflict of interest None.

Ethics approval None.

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