Letter to the Editor

Ectopic double primary bronchogenic cysts: a case description

Xinlong Liu[1,2]^, Teng Sun[1,2]^, Tao Hong[1,2], Hao Zhang[1,2]^

[1]Thoracic Surgery Laboratory, Xuzhou Medical University, Xuzhou, China; [2]Department of Thoracic Surgery, Affiliated Hospital of Xuzhou Medical University, Xuzhou, China

Correspondence to: Hao Zhang, MD, PhD. Department of Thoracic Surgery, Affiliated Hospital of Xuzhou Medical University, 99 West Huaihai Road, Xuzhou 221006, Jiangsu, China; Thoracic Surgery Laboratory, Xuzhou Medical University, Xuzhou 221006, China. Email: zhanghao@xzhmu.edu.cn.

Submitted Feb 28, 2023. Accepted for publication Jul 06, 2023. Published online Aug 24, 2023.
doi: 10.21037/qims-23-247
View this article at: https://dx.doi.org/10.21037/qims-23-247

Introduction

Bronchogenic cyst (BC) is a rare congenital developmental abnormality, usually affecting the lungs and mediastinum. BCs occurring at abnormal sites such as the neck, abdominal cavity, retroperitoneum, and spinal dura are extremely rare and are referred to as ectopic bronchogenic cysts (eBCs). BCs are asymptomatic and lack typical imaging findings in the initial stage; however, as the cyst grows, it compresses the surrounding tissues and organs, causing clinical symptoms, which are not recognized until the occurrence of complications such as secondary acute bleeding, infection, and perforation. Therefore, most patients cannot receive timely treatment at the early stage of the disease, resulting in aggravated disease and poor prognosis. Similar to single BCs, ectopic double primary BCs have no clinical symptoms at the initial stage and are not typical in imaging. Early diagnosis still faces great challenges, as ectopic double primary BCs are easily confused with neurogenic tumors, diaphragmatic hernias, hydatid cysts, esophageal diverticulum, or adrenal cysts. Computed tomography (CT) and magnetic resonance imaging (MRI) are still the best imaging methods for BCs.

The mechanism of BC emergence remains unclear; however, the case we report here strongly supports Sumiyoshi *et al.*'s hypothesis of germ shedding and displacement (1) and provides case evidence for further defining the pathogenesis of BCs. In addition, most of the reported cases of BCs have been found by chance without clinical symptoms or complications. Based on the unique medical experience of a patient we treated, we witnessed the whole process from asymptomatic diagnosis to the appearance and gradual worsening of symptoms. This closely observed course of clinical symptom evolution provides principle clinical evidence for the early diagnosis and treatment of BCs.

Case presentation

During a physical examination, a retroperitoneal and mediastinal mass was incidentally detected in a 54-year-old middle-aged male. He had no symptoms of discomfort in his daily life, and laboratory and physical examination results of the chest and abdomen were normal. CT showed a circular high-density shadow with clear boundary on the left side of the T11 vertebral body, approximately 31×23 mm^2 in size. The cyst exhibited moderate enhancement upon administration of contrast agent. Furthermore, an irregular mass shadow at the bottom of the left side of the diaphragm was observed, the boundary with the left adrenal gland was not clear, and the adrenal gland was compressed. The maximum cross-section area was about 47×32 mm^2, and multiple punctate and lamellar calcifications were observed at the edge of the lesion, but the enhancement was not obvious (*Figure 1*). As the boundary between the tumor and the patient's adrenal gland was not clear and because the adrenal gland was compressed, surgical treatment was recommended by the urologist. Retroperitoneal laparoscopic resection was performed in May 2019. Intraoperative exploration revealed

^ ORCID: Xinlong Liu, 0000-0002-1860-5767; Teng Sun, 0000-0002-9125-5070; Hao Zhang, 0000-0002-2926-737X.

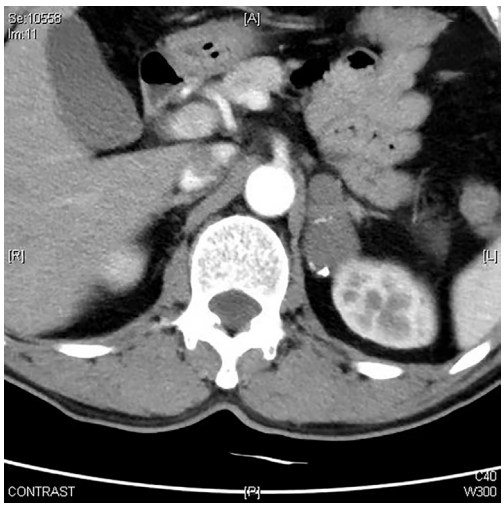

Figure 1 Imaging findings of bronchogenic cysts. An irregular mass shadow could be seen at the left diaphragmatic crura, the boundary with the left adrenal gland was not clear, and the left adrenal gland was compressed.

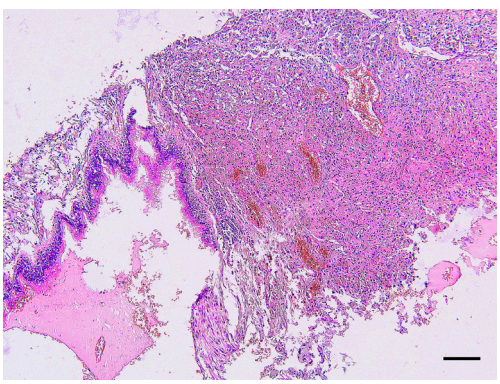

Figure 2 Retroperitoneal laparoscopic resection of the tumor (HE staining). Ciliated columnar epithelium and adrenal tissue were observed under microscopy. Bar = 100 μm. HE, hematoxylin and eosin.

that the tumor was present at the ventral side and was approximately 50×40×20 mm^3 in size, with a grayish yellow color and a false capsule. The surgeon exposed the tumor from the adrenal gland to the ventral side along the upper polar surface of the kidney and resected it completely. The abdomen was closed after the placement of drainage tubes. The resected specimen was a piece of solid gray–red tissue, approximately 80×50×20 mm^3 in size. The size of the solid part was approximately 50×45×17 mm^3. The section of the solid area was gray-white and gray-red, jelly-like, and soft, and part of adrenal tissue could be seen in the section (*Figure 2*). The cyst was 20×15×10 mm^3 in size, and the wall of the capsule had been damaged, with some grey–brown jelly residue remaining. Pathological analysis showed that the cyst had typical broncho-ciliated columnar epithelial tissue, and it was thus considered to be an eBC (*Figure 3A*,*3B*).

One year later, the doctor at the thoracic surgery clinic of Xuzhou Medical University Affiliated Hospital recommended surgery, but the patient refused due to discomfort and concerned regarding the risk of surgery. Half a year later, he developed a symptom of back pain, which gradually worsened. He was thus hospitalized at the Department of Thoracic Surgery, Affiliated Hospital of Xuzhou Medical University. Reexamination CT showed a circular low-enhancement shadow on the left side of the T11 vertebral body with a clear boundary, approximately 31×21×10 mm^3 in size, and an arc-shaped calcification at the local edge. There was no significant change compared with previous CT results. For further diagnosis, the patient underwent MRI, which showed a circular, long T2, and a long T1 signal shadow on the left side at the T11 level (*Figure 4*), indicating the presence of a cystic mass. Based on the patient's medical history, we considered the possibility of BCs and performed robot-assisted thoracoscopic mediastinal mass resection. During the operation, a mass in the posterior mediastinum, approximately 30×30×10 mm^3 in size, was identified with a complete, round, and soft capsule. The surgeon removed the tumor tissue completely after removing the surrounding tissue. The resected specimen was a gray-red cystic mass, approximately 38×20×10 mm^3 in size; the section surface was multilocular cystic, containing a grayish brown jelly-like material; the cyst wall was 1- to 2-mm thick, and the inner wall was smooth. Pathological analysis showed that the cyst had typical bronchial tissue: cartilaginous tissue and ciliated columnar epithelial tissue. It was thus considered a mediastinal BC (*Figure 5A*,*5B*). After the operation, the patient recovered smoothly, and the clinical symptoms were significantly relieved. After discharge, the patient was followed up regularly (*Figure 6*). No recurrence or discomfort was observed for more than 2 years postsurgery.

All procedures described in this study were performed in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Declaration of Helsinki (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images.

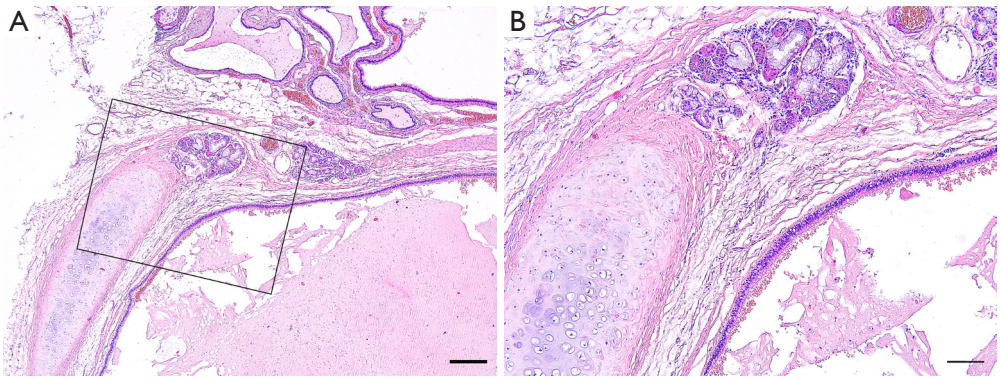

Figure 3 Microscopic features of bronchogenic cysts (HE staining). (A) Pathological section of the rBC. Bar = 300 μm. (B) Typical bronchial structure including cartilaginous tissue, a ciliated columnar epithelium, and surrounding small salivary glands. Bar = 100 μm. HE, hematoxylin and eosin; rBC, retroperitoneal bronchogenic cyst.

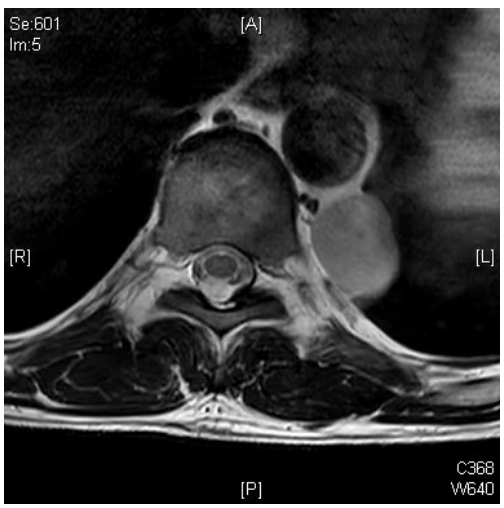

Figure 4 Imaging findings of bronchial cysts. At the left side of the T10 level, a circular long T2 and long T1 signal shadow were observed.

A copy of the written consent is available for review by the editorial office of this journal.

Discussion

A BC is an abnormal developmental anomaly of the foregut, usually occurring in the mediastinum. According to the occurrence site, BCs can be divided into mediastinal, intrapulmonary, and ectopic types. The ectopic type is rare and has been reported in the neck, subcutaneous tissue, atrium, spinal canal, abdominal cavity, retroperitoneum, and spinal dura (2-6). In the case described here, the patient had 2 BCs at 2 sites: 1 above and 1 below the diaphragm. The subdiaphragmatic cyst was a retroperitoneal BC (rBC), which is extremely rare. This is the first case report on ectopic double primary BCs.

To date, the mechanism of BCs remains unclear. In 1985, Sumiyoshi *et al.* (1) proposed the germ-shedding-and-displacement hypothesis, which states that during embryonic development, with the formation of diaphragm, the pericardioperitoneal canal separates to form the chest and abdominal cavity (7,8). The original bronchial tree undergoes shedding and migration for various abnormal reasons, the embryo is shed and transferred to the caudal side, and a BC is formed at the migration site. Another hypothesis has been proposed that BCs arise from abnormal differentiation of a foregut-derived cyst in the abdominal cavity (6,9), which explains the possibility of it occurring in the liver, pancreas, stomach, and other organs (*Table 1*). In the present case, the locations of the 2 BCs in the patient were close to each other, separated only by the diaphragm. We speculated that the detached and migrated germ was separated by the diaphragm during the development of the patient's chest and abdomen, resulting in the formation of 2 cysts—one in the chest and the other in the abdomen—which provides strong evidence supporting the hypothesis of Sumiyoshi *et al.* (1).

BCs are usually asymptomatic at the initial stage because the disease develops relatively slowly (6,10,11). However, as the cyst grows, it compresses the surrounding tissues and organs, causing related clinical symptoms, which are not recognized until the occurrence of complications such as secondary acute bleeding, infection, perforation, and malignancy (12-16). Therefore, most patients do not receive

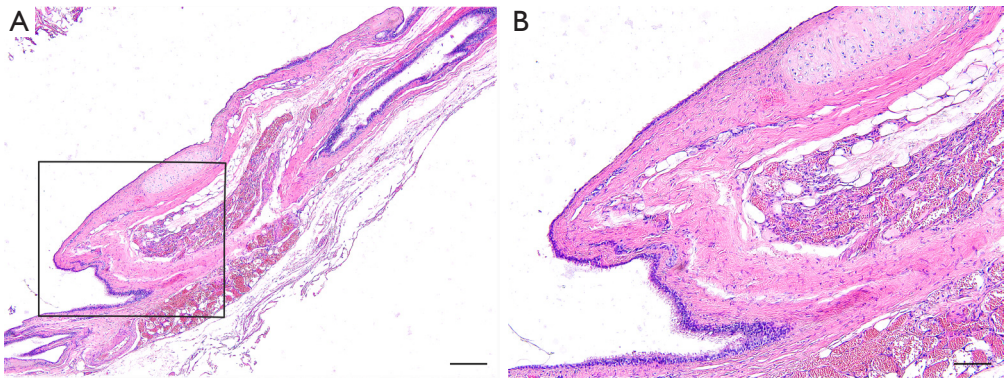

Figure 5 Microscopic features of the bronchogenic cysts (HE staining). (A) Pathological section of mediastinal bronchogenic cysts. Bar = 300 μm. (B) Typical bronchial structure including cartilaginous tissue and a ciliated columnar epithelium. Bar = 100 μm. HE, hematoxylin and eosin.

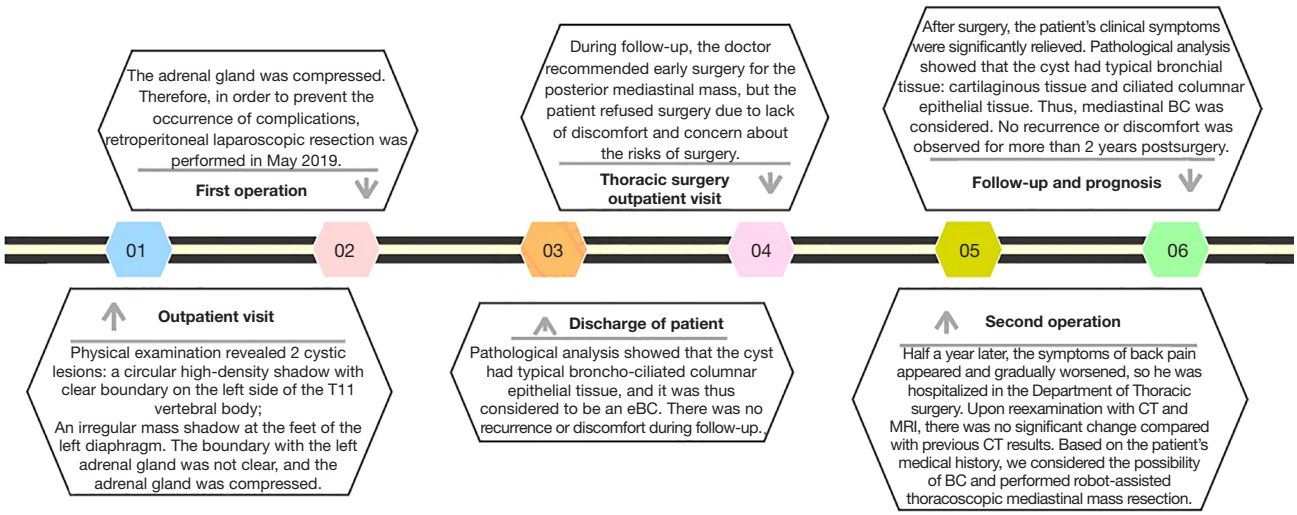

Figure 6 The treatment course of the patient's 2 cysts from the physical examination to the end of treatment. CT, computed tomography; MRI, magnetic resonance imaging.

timely treatment in the early stage of the disease. In this case, the patient's rBC was accidentally discovered during physical examination. Owing to the slow growth of the cyst and the relatively large retroperitoneal space, no obvious discomfort was experienced by the patient. This case is consistent with the genesis and clinical manifestations of BCs in most patients (1,8,17,18). However, in this case, the mediastinal BCs of the patient evolved from no clinical symptoms at the time of initial discovery to back pain; from the initial refusal of surgery to the final surgical treatment, we witnessed the progression in detail. Based on the unique medical experience of this patient, we suggest that for BCs, complete resection of the cyst should be performed immediately to prevent the occurrence of clinical symptoms, related complications, and even the possibility of malignancy, even if there are no related symptoms or complications at the initial diagnosis.

CT and MRI are the best imaging methods for the diagnosis of BCs (10,11,19,20), but the imaging findings are not typical (21). CT usually presents low-density circular cystic lesions, which are mostly not enhanced by enhanced scanning. Due to the presence of protein in the cysts or their being accompanied by infection and bleeding, the CT value of some lesions is increased; thus, BCs can be easily

Table 1 Common sites and first clinical symptoms of ectopic bronchogenic cysts reported in the literature

Author	Publication year	Cyst site	Cyst number	Clinical symptoms
Sumiyoshi *et al.*	1985	Retroperitoneum adjacent to the pancreatic	1	Epigastric pain accompanied by nausea and vomiting
Ali *et al.*	2018	Subcarinal mass	1	Severe back pain, epigastric distress
Bai *et al.*	2021	Left atrium	1	Intermittent chest pain
Flórez Rial *et al.*	2022	Pancreas	1	No clinical symptoms
Wen *et al.*	2020	Adrenal gland area	1	No clinical symptoms
Liang *et al.*	2005	Subdiaphragmatic	1	Intermittent left flank pain
Tang *et al.*	2021	Diaphragm	1	Coughs and expectoration
Erbenová *et al.*	2021	Gastric cardia wall	1	No clinical symptoms
Kim *et al.*	2021	Presacral space	1	No clinical symptoms

confused with neurogenic tumors, diaphragmatic hernias, hydatid cysts, or esophageal diverticulum (20). MRI usually shows low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Owing to the contents of the cyst, fat suppression in the T1-weighted image cannot be observed for BCs, which can otherwise be used to distinguish teratoma from dermoid cyst (19). The final diagnosis of BCs depends on pathological examination. BCs has the following pathological features: a ciliated columnar epithelium, a mucous gland, smooth muscle fiber, fibrous tissue, elastic tissue, and cartilage (7-8,17,21). In our case, ciliated columnar epithelial structures were observed in both pathological sections of the patient, which confirmed the diagnosis of BCs.

In the treatment of BCs, total surgical resection should be considered first to avoid recurrence of the disease due to incomplete resection of the cyst wall (13,19,20,22). The location of BCs is variable, so the difficulty of operation varies greatly. We have the following experience in the treatment: First, when the cyst is not large, or the adhesion is not serious, the surrounding tissue should be separated to ensure the complete resection of the cyst wall without residual material. Second, when the cyst is large, or there are severe adhesions, the volume can be reduced to facilitate the operation and finally complete resection of the cyst wall. Third, cysts occurring in specific locations need to be analyzed case by case to maximize the patient's benefit. For example, in our case, in which the patient had a BC in the carina of the sac wall, which is closely related to the tracheal membrane, the surgeon had 2 choices: protect the tracheal membrane and preserve part of the sac wall—which entailed a risk of malignant transformation of the residual sac wall and recurrence—or complete dissection of the cyst—which might have damaged the tracheal membrane. Damage to the tracheal membrane could not be completely avoided; thus, whether the treatment and repair should have been carried out was also a difficult decision for the surgeon. Based on the long-term benefit of the patient and confidence in his own level of performing the operation, the surgeon finally separated the sac wall from the tracheal membrane meticulously and achieved the most beneficial result for the patient.

Conclusions

In the present case of ectopic double primary BCs, the 2 BCs were located in close proximity to the chest and abdomen and separated by the diaphragm, which strongly supports Sumiyoshi *et al.*'s hypothesis of germ shedding and displacement (1). This report thus provides crucial evidence for further defining the pathogenesis of BCs.

In this case, the entire process of diagnosis and treatment of mediastinal BCs was fully observed from asymptomatic diagnosis to the appearance and gradual worsening of symptoms. Thus, this report may provide a clinically based recommendation for the diagnosis and treatment of BCs, namely, early surgical treatment after discovery.

Minimally invasive surgery (including thoracoscopy and robotics) is an ideal method for the treatment of BCs. For patients with multiple BCs, simultaneous or sequential

surgery can be performed.

Acknowledgments

Funding: The study was supported by The Social Development Projects of Key R&D Programs in Xuzhou City (No. KC22097).

Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-23-247/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Declaration of Helsinki (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

References

1. Sumiyoshi K, Shimizu S, Enjoji M, et al. Bronchogenic cyst in the abdomen. Virchows Arch A Pathol Anat Histopathol 1985;408:93-8.
2. Ali S, Rauf A, Meng LB, et al. Case Report: Severe back pain, epigastric distress and refractory nausea; an unusual presentation of mediastinal bronchogenic cyst. F1000Res 2018;7:960.
3. Gui Z, Cai T, Tian Y, et al. Intrathyroidal Bronchogenic Cysts: A report of two cases. J Int Med Res 2022;50:3000605221087032.
4. Bai W, Tang H. A giant bronchogenic cyst compressing the left atrium: A case report. Anatol J Cardiol 2021;25:E11-2.
5. Kweh BTS, Roberts-Thomson S, Verhellen T, et al. Bilobed intramedullary and extramedullary bronchogenic cyst of the conus medullaris: illustrative case. J Neurosurg Case Lessons 2021;2:CASE21323.
6. Flórez Rial MDP, Ruz Portero S, Valenzuela González M, et al. Intrapancreatic bronchogenic cyst. A case report. Rev Esp Patol 2022;55:292-6.
7. Wang E, Zhang L, Wang Y, et al. Retroperitoneal bronchogenic cyst adjacent to aortaventralis: A case report. Asian J Surg 2023;46:1583-4.
8. Wen Y, Chen W, Chen J, et al. Retroperitoneal bronchogenic cyst resembling an adrenal tumor: two case reports and literature review. J Int Med Res 2020;48:300060520925673.
9. Liang MK, Yee HT, Song JW, et al. Subdiaphragmatic bronchogenic cysts: a comprehensive review of the literature. Am Surg 2005;71:1034-41.
10. Tang J, Zeng Z, Deng S, et al. Ectopic bronchogenic cyst arising from the diaphragm: a rare case report and literature review. BMC Surg 2021;21:321.
11. Erbenová A, Placrová B, Špůrková Z, et al. Bronchogenic cyst of gastric cardia - case report and literature review. Rozhl Chir 2021;100:507-11.
12. Han KZ, Tuerhong A, Wang SP, et al. Retroperitoneal Bronchogenic Cyst: A Case Report. Journal of Hepatopancreatobiliary Surgery 2019;31:571-2.
13. Gong ZW, Yao XX, You GQ, et al. A case of retroperitoneal bronchogenic cyst. Journal of Clinical Hepatology 2018,34:616-7.
14. Hu BY, Yu H, Shen J. A retroperitoneal bronchogenic cyst clinically mimicking an adrenal mass: three case reports and a literature review. J Int Med Res 2022;50:3000605211072664.
15. Gutiérrez GS, Gutiérrez FG, Bastianelli GA, et al. Bronchogenic cyst in an unusual location. Asian Cardiovasc Thorac Ann 2021;29:44-6.
16. Matsushita M, Honda O, Kohzai M, et al. Bronchogenic cyst with atypical imaging findings and repeated ruptures in a short period of time: A case report. Radiol Case Rep 2022;17:3933-7.
17. Runge T, Blank A, Schäfer SC, et al. A retroperitoneal bronchogenic cyst mimicking a pancreatic or adrenal mass. Case Rep Gastroenterol 2013;7:428-32.
18. Pasquer A, Djeudji F, Hervieu V, et al. A rare retrorectal presentation of a bronchogenic cyst: A case report. Int J

Surg Case Rep 2016;24:112-4.
19. Wang M, He X, Qiu X, et al. Retroperitoneal bronchogenic cyst resembling an adrenal tumor with high levels of serum carbohydrate antigen 19-9: A case report. Medicine (Baltimore) 2017;96:e7678.
20. Chung JM, Jung MJ, Lee W, et al. Retroperitoneal bronchogenic cyst presenting as adrenal tumor in adult successfully treated with retroperitoneal laparoscopic surgery. Urology 2009;73:442.e13-5.
21. Kim AY, Min SJ, Kim H, et al. Retroperitoneal Bronchogenic Cyst Located in the Presacral Space: A Case Report. Taehan Yongsang Uihakhoe Chi 2021;82:207-11.
22. Fievet L, Gossot D, de Lesquen H, et al. Resection of Bronchogenic Cysts in Symptomatic Versus Asymptomatic Patients: An Outcome Analysis. Ann Thorac Surg 2021;112:1553-8.

Cite this article as: Liu X, Sun T, Hong T, Zhang H. Ectopic double primary bronchogenic cysts: a case description. Quant Imaging Med Surg 2023;13(9):6310-6316. doi: 10.21037/qims-23-247

