CASE REPORT

Bronchogenic cyst arising from the crus of the left hemidiaphragm

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SUMMARY

A 34-year-old man presented with chronic worsening left-sided retrosternal chest pain. Following a negative cardiac work up he was found, on cross-sectional imaging, to have a cystic mass measuring 9.6×11.8×9 cm related to his left diaphragmatic crus. The patient underwent an exploratory laparotomy with complete resection of the cystic mass. Histopathological examination of the mass confirmed it as being a bronchogenic cyst. His pain resolved following excision of the mass and at follow-up he was asymptomatic with no evidence of recurrence on imaging.

BACKGROUND

Bronchogenic cysts are rare congenital lesions that arise from the ventral diverticulum of the primitive foregut¹ after the third week of embryonic development. In most cases, the cysts are associated with the tracheobronchial tree (although lacking continuity)² and are most commonly found in the posterior carina, however, they may also be found in atypical locations, such as intraspinal,³ subdiaphragmatic or retroperitoneal regions.¹ To date, there have been 48 cases of subdiaphragmatic bronchogenic cysts described in the English literature, with retroperitoneal location being very rare.⁴ Bronchogenic cysts have been described in various ages and genders with a female predominance of 65%.^{5,6}

Most cases are asymptomatic, with the majority of bronchogenic cysts discovered incidentally after imaging such as CT or MRI, where they are often misdiagnosed given their non-specific imaging features.⁷ The main differential diagnoses include teratoma, abscess, lymphocele and neoplasm.⁷ Cysts usually become symptomatic after rupture, secondary infection of the cyst or compression of surrounding structures.^{3,7} When present, the most common symptoms include worsening pain, dyspnoea, cough and dysphagia.⁸ Histopathological assessments usually reveal the presence of bronchial tissue within the cysts,² having ciliated pseudostratified columnar epithelium with the wall containing smooth muscle, cartilage, bronchial glands or linear calcifications.⁶ Therefore, histopathology is used to obtain a definitive diagnosis.

Surgical resection remains the treatment modality of choice due to its ability to provide a tissue sample for a definitive diagnosis as well as providing a cure if the cyst is resectable. Resections for cysts located adjacent to major organs such as the pancreas, adrenals or the diaphragm make the resection more challenging.⁷ Following excision, as

long as a complete excision has been performed, recurrence is low and prognosis excellent.

CASE PRESENTATION

A 34-year-old man presented to an outside hospital, with left-sided chest pain. The pain was described as constant, dull and located just inferior to the sternum and radiated to his back. The patient also noted that the pain had been increasing in severity over the past 3 months. It was alleviated by sitting up and exacerbated by lying down. The patient denied shortness of breath, nausea, vomiting, change in bowel habit, fever or chills. His medical history included asthma.

On physical examination, he was found to be in no acute distress, the lungs were clear to auscultation bilaterally with good diaphragmatic excursion and cardiac examination was normal. The abdomen was soft and non-distended with mild tenderness in the superior epigastrium without rebound. No mass was palpable.

Vital signs were normal with a heart rate of 84 bpm, blood pressure of 140/88 mm Hg, temperature of 36.6°C, respiratory rate of 20 breaths/min and oxygen saturation of 97% on room air.

INVESTIGATIONS

Cardiac workup was found to be negative for ischaemic heart disease. A CT scan of the chest

Figure 1 CT of the abdomen showing a 9.7×10.9×8.3 cm well encapsulated mass of decreased attenuation that is partially calcified, arising primarily from the diaphragmatic crus of the left hemidiaphragm caudal to the gastro-oesophageal junction.

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Figure 2 MRI demonstrating a cystic mass with a few posterior septations abutting the left diaphragmatic crus.

revealed a well-encapsulated mass measuring $9.7 \times 10.9 \times 8.3$ cm. The mass was of decreased attenuation and partially calcified and arose from the crus of the left hemidiaphragm caudal to the gastro-oesophageal junction. The mass was located medial to the left lesser curve of the stomach and superior to the body of the pancreas (figure 1).

The patient subsequently had a basic set of laboratory tests drawn showing white cell count $6.57 \text{ k}/\mu\text{L}$, haemoglobin $13.5 \text{ g}/\text{dL}$, hematocrit 42.4% , platelets $212 \text{ k}/\mu\text{L}$, Na $141 \text{ mmol}/\text{L}$, K $3.8 \text{ mmol}/\text{L}$ and Cr $1.24 \text{ mg}/\text{dL}$. Troponin was found to be $<0.01 \text{ ng}/\text{mL}$ with creatine kinase of $583 \text{ U}/\text{L}$.

To further delineate the mass, an MRI scan of the chest was performed and confirmed the presence of a cystic retroperitoneal mass without enhancing components (figure 2).

DIFFERENTIAL DIAGNOSIS

Possible diagnoses included lymphoma, pancreatic pseudocyst, pancreatic serous cystadenoma, duplication or inclusion cyst and lymphangioma.

TREATMENT

After informed consent was obtained, the patient underwent an exploratory laparotomy. Intraoperatively, the retroperitoneal

mass was found to be arising on a pedicle from the left diaphragmatic with abutment but no invasion of the pancreas and stomach. The mass was excised and histopathological evaluation revealed a bronchogenic cyst with chronic inflammation, granulation tissue formation and dystrophic calcification but no evidence of neoplasia (figure 3).

OUTCOME AND FOLLOW-UP

The patient tolerated the procedure well and was discharged on postoperative day 4. On the day of discharge, the patient was able to tolerate a full diet and only endorsed the expected per-incisional tenderness. He made an uneventful recovery from his surgery and his pain resolved over the first 2 postoperative months. At 10 months follow-up, his symptoms had completely resolved.

DISCUSSION

Bronchogenic cysts are rare congenital abnormalities arising from the primitive foregut after the third week of development. As these cysts are associated with the tracheobronchial tree, the majority of cysts are found in the mediastinum. Rarely, bronchogenic cysts may be found subdiaphragmatically when the abnormal buds migrate inferiorly prior to the fusion of the diaphragm.^{1 7} Although the majority of such cases are located in the left retroperitoneal region, retroperitoneal bronchogenic cysts still remain a very rare occurrence. The most common location for retroperitoneal lesions, where $>80\%$ of such cysts have been found, is in the following triangle: posterior portion of the stomach, splenic vein and spleen/diaphragm.⁴

Bronchogenic cysts are often incidental findings discovered on imaging, most commonly CT or MRI. Oftentimes, retroperitoneal cysts are asymptomatic, given the significant amount of potential space for growth.⁷ Interestingly, the largest bronchogenic cyst described to date was found to be 18 cm in diameter.⁹

CT imaging remains the first step in working up the patient. The most common finding on CT imaging for cyst lesions is either a spherical mass with water or a spherical mass with soft tissue attenuation.⁸ When the attenuation resembles soft tissue, bronchogenic cysts are more difficult to differentiate from other mediastinal or intra-abdominal processes (depending on the location of the cyst). Therefore, MRI and ultrasound (US) are usually both needed for better evaluation.⁸ However, the utility of US is limited due to the deep location of the cyst in many patients.⁷

The treatment of choice remains surgical excision, as it provides a tissue sample for a definitive diagnosis as well as resolution of symptoms in cysts causing nearby structure

Figure 3 (A) Fibromuscular cyst wall with chronic inflammation lined by respiratory-type epithelial lining consisting of pseudostratified epithelial cells with terminal bar and cilia (H&E, original magnification $\times 40$ and $\times 400$ (inset)). (B) Focal areas with dystrophic calcifications and granulation tissue, respiratory-type epithelium present on the left (H&E, original magnification $\times 100$).

compression. Even in the case of initially asymptomatic bronchogenic cysts, surgical resection is recommended, given the risks of malignant transformation in 0.7% of cases as well as eventual symptom development.¹⁰ The malignant transformations observed have included adenocarcinoma, squamous cell, bronchoalveolar carcinoma and large cell anaplastic carcinoma.¹⁰ Therefore, long term close follow-up is needed if the cysts are not removed.¹⁰

Learning points

- ▶ Bronchogenic cysts arise after the third week of development during abnormal budding from primitive foregut. Most are located in the mediastinum with retroperitoneal location being very uncommon.
- ▶ Histopathology is required for establishment of a definitive diagnosis.
- ▶ Surgical resection remains the treatment modality of choice, given 0.7% malignant potential as well as eventual symptom development in 50% cases.

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Competing interests None declared.

Patient consent Obtained.

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