Cardiology in the Young

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Bronchogenic cyst presenting with respiratory distress in a neonate

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Original Article

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Abstract

Bronchogenic cysts are considered rare airway anomalies that can have a significant impact on the well-being of infants. In this case report, we present a rare presentation of bronchogenic cyst presenting with early neonatal respiratory distress due to airway and vascular compression. Surgical excision was curative with an excellent prognosis.

Bronchogenic cysts are considered part of rare airway anomalies that can have a significant impact on the well-being of infants. Embryologically, they comprise part of a spectrum of *foregut duplication cysts*, with the remaining two types being oesophageal duplication cysts and enteric duplication cysts. Bronchogenic cysts occur typically in the middle mediastinum and comprise around 10% of all mediastinal masses and around 50% of all mediastinal cysts.[1,2] The involved site of bronchogenic cysts depends on the timing of the malformation during embryonic development. In the early embryonic development, budding malformation resulting in a bronchogenic cyst is confined to pulmonary and thoracic locations, whereas later development of the malformation can involve more distant tissues such as the oral cavity, oesophagus, and abdomen. Intracardiac and pericardial involvement is also described. Differential diagnosis includes oesophageal or enteric duplication cysts, pericardial cysts, cystic hygromas, and lymphangiomas.[3]

Although many reported patients were asymptomatic, neonatal presentation can be associated with respiratory and/or feeding difficulties. Antenatal detection has become more prevalent and is estimated to detect around 60% of cases of bronchogenic cysts.[4]

We report a neonate who presented with respiratory distress and was found to have a huge mediastinal mass which was confirmed to be a bronchogenic cyst on histopathological examination.

Case presentation

A 29-day-old female neonate was referred to King Abdulaziz Cardiac Center (KACC) because of respiratory distress. She was a product of a caesarean section after a full-term uneventful pregnancy in Hail General Hospital. Her mother was healthy with one previous termination of pregnancy and no living children. She developed severe respiratory distress immediately after birth, required intubation and ventilation, and received two doses of synthetic surfactant. She was extubated at 11 days of age and then transferred to the general ward. Her septic workup was negative. However, she remained in respiratory distress needing supplemental oxygen, so a transthoracic echocardiogram was performed and showed a huge cystic mass compressing the surrounding structures. Subsequently, she was referred to our cardiac centre.

On arrival to our Pediatric Cardiology Ward, her weight was 2.9 kg, pulse 146 per minute, and respiratory rate 48 per minute. She was in notable distress with tachypnoea, intermittent flaring, and grunting. However, no stridor or wheezing was noted. She was maintaining normal oxygen saturation on minimal oxygen by nasal cannula. Her chest radiograph did not show a major abnormality. Her echocardiogram (Fig 1) showed a large cystic chest mass originating posteriorly and exerting compression effects on the left atrium and pulmonary artery branches. The right pulmonary artery was severely compressed with diminished flow and high peak gradient, while the left was less affected. The mass appeared to have no clear communication with the heart, which was confirmed by CT imaging (Fig 2). For a rotational movie of rendered 3-d images, please refer to the data supplement (Movie 1).

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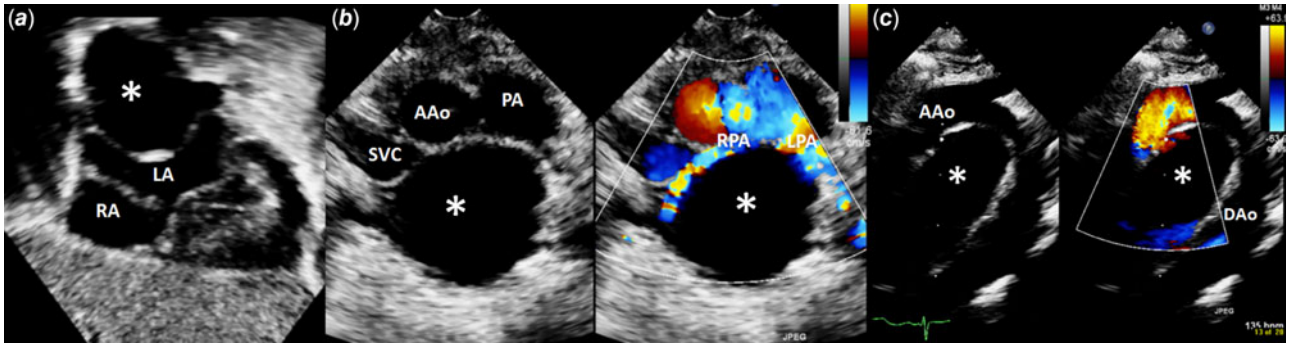


Figure 1. Transthoracic echocardiographic images. Transthoracic echocardiographic views showing a huge mass posterior and superior to the heart with compression of the left atrium, as seen from subcostal (***a***), high parasternal (***b***), and suprasternal views (***c***). The Color Flow Doppler (***b***, on the right) demonstrates branch pulmonary arteries compression. The mass was extending superiorly underneath the aortic arch (***c***). No communication of flow with the mass was noted on Color Doppler Flow (***b***, ***c***).

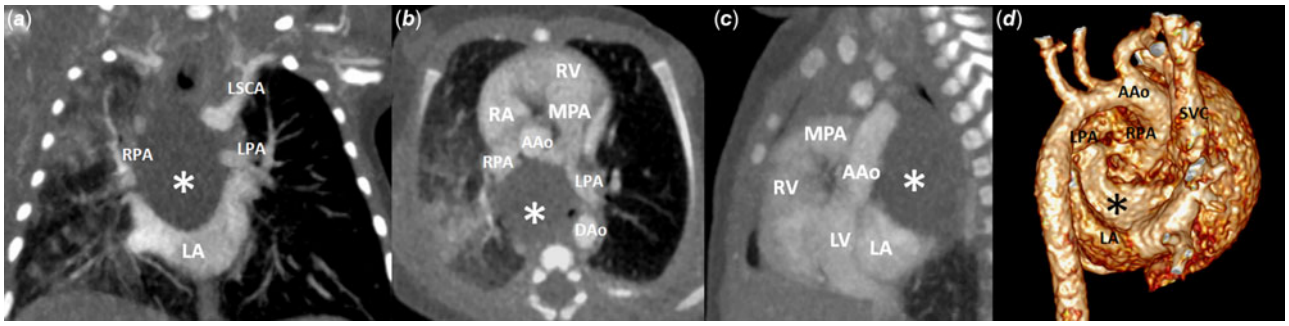


Figure 2. CT images. CT angiographic coronal (***a***), axial (***b***) sagittal, (***c***), and 3D (***d***) views demonstrating a huge extra-cardiac nonenhanced homogeneous mass, with fluid content based on the Hounsfield unit. The mass measures 21 × 23 mm and is located in the superior–posterior aspect of the left atrium, compressing the airway and pulmonary artery branches, and shifting the aortic arch towards the left side. No communication was seen between the heart structures and this mass. It appears as a space-occupying lesion in the maximum intensity projection and 3D views.

The patient underwent surgery on the 25 June, 2020. The surgery was performed by a cardiac surgeon and a paediatric surgeon together with standby cardiopulmonary bypass, via a median sternotomy. A large cystic mass was found superior to the left atrium (Fig 3a). It originated from the trachea just proximal to the bifurcation. It was tense and compressing the surrounding structures including the aorta and pulmonary arteries. The mass was mobilised free from all surrounding structures and resected off the trachea with no tracheal extension. The cyst was full of a clear serous fluid which leaked during excision, and the mass was sent for histopathological evaluation after resection. Intraoperative transoesophageal echocardiography showed no significant residual lesion.

The histopathological examination (Fig 3b) showed the cyst to be unilocular and lined by ciliated pseudostratified columnar epithelium of respiratory type. The wall contained hyaline cartilage, smooth muscle, bronchial glands, brown fat, and nerve fibres. Those findings were consistent with a diagnosis of a bronchogenic cyst.

After surgery, she was transferred to the Pediatric Cardiac Intensive Care Unit (PCICU) and successfully extubated within 24 hours. She developed left lung atelectasis that was found to be related to left diaphragmatic paresis confirmed by ultrasound examination. Most probably this was due to left phrenic nerve paresis which was close to the lesion during dissection. She had transient hypertension which was controlled by medication. Feeding was established and well tolerated. She stayed 10 days in the Paediatric Cardiac Intensive Care Unit after which she was transferred to the Stepdown Unit in a stable condition. Oxygen was weaned off gradually and feeding was optimised. She was discharged home in a healthy condition after 20 days of total hospital stay. At the follow-up visit, the infant was asymptomatic and had no residual lesion on echocardiography.

Discussion

Bronchogenic cysts are rare congenital malformations with an estimated incidence of 1 in every 42,000–68,000 newborns and constitute 6–10% of mediastinal masses in children.[5] Bronchogenic cysts develop from unusual airway budding and have been described in relation to the airway as paratracheal, intraluminal, or intramural.[6] The differential diagnosis for other congenital cystic lung lesions includes congenital cystic adenomatoid malformation, pulmonary sequestration, and congenital lobar emphysema.[6] Antenatal detection has been reported with increased frequency since the wide application of routine antenatal ultrasound and bronchogenic cysts appear usually as an anechoic image.[7] Postnatal symptomatic presentation is rare, however, when present (as we have seen in our patient), it may result in a significant compression effect on the surrounding structures warranting an urgent surgical excision.[2] Mortality has been described due to severe tracheal obstruction, and cardiac arrest has been described in the literature.[8] Some reports have linked bronchogenic cysts with the risk of developing malignant transformation.[9] Further imaging by CT or MRI is recommended before surgical excision to further delineate the mass and differentiate it from surrounding tissues.[3] Endoscopic ultrasonography is a valuable tool for the evaluation of such masses, and the characteristic features are described in several publications

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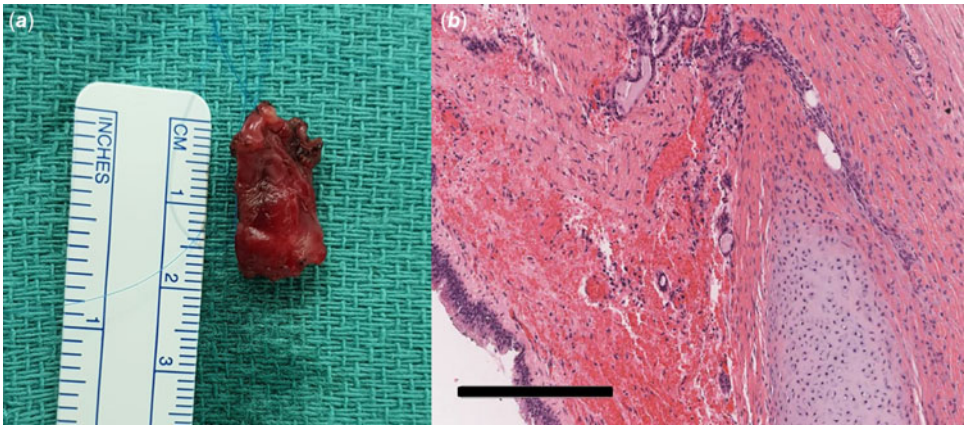


Figure 3. Gross anatomy and histopathology. (***a***) Gross appearance of the surgically excised cystic mass, measuring around 18 × 8 mm. (***b***) Histological section showing the cyst wall with a lining of respiratory epithelium*, bronchial glands**, and cartilage*** (H&E stain). Bar = 0.300 mm.

in the adult population.[10] Bronchoscopy is helpful to delineate the communication of the cyst with the trachea and whether tracheal surgery is needed or not.[11]

The histological differential diagnosis includes cystic teratoma and oesophageal cyst. The cystic teratoma contains tissue derived from all three germ cell layers and lined by variable mixtures of gastrointestinal, squamous, and respiratory epithelium and may contain immature tissues as well. In comparison, oesophageal cysts are usually attached to the oesophageal wall lined by stratified squamous or gastrointestinal epithelium and contain a double smooth muscle layer.[12]

The natural history of bronchogenic cysts was demonstrated by Maurin et al., in a study involving six patients who were followed from the antenatal period till the time of surgical excision.[13] These authors concluded that cysts have exponential growth from about one to two centimeters at birth to ten centimeters in adulthood, and subsequently, excision before age of 2 years is recommended. In a large series published by Kieran et al., including 22 patients, surgical excision was shown to be curative with no instances of recurrence.[2]

Conclusions

We highlight the importance of including this rare entity in the differential diagnosis of mediastinal masses with airway and vascular compression in neonates. Surgical excision with complete removal of the mucosal lining is curative, with no residual lesion seen on follow-up.

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