

Bronchogenic Cyst in the Interatrial Septum with a Single Persistent Left Superior Vena Cava

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Bronchogenic cysts are classified as a type of congenital foregut cyst. They are usually extrapulmonary and occur mostly along the tracheobronchial tree. Intracardiac bronchogenic cysts are very rare. As far as we know, only 3 cases have been reported previously. This paper presents a unique case of a 70-year-old male who was diagnosed with a bronchogenic cyst in the interatrial septum with a concomitant persistent left superior vena cava. He underwent successful complete excision of the cyst under cardiopulmonary bypass and enjoyed an uneventful life with normal sinus rhythm during the 1-year follow-up. The relevant literature is reviewed and an explanation for the embryogenesis of the bronchogenic cyst in such a peculiar location is proposed. [*J Chin Med Assoc* 2006;69(2):89–91]

Key Words: atrial fibrillation, bronchogenic cyst, heart atria, heart septum, superior vena cava

Introduction

Bronchogenic cysts are considered as congenital anomalies that originate from the ventral foregut during embryologic development. They are the most common primary cysts of the mediastinum, although they can also be found in the lung parenchyma. However, intracardiac bronchogenic cysts are very rare. This paper describes a unique case of a bronchogenic cyst in the interatrial septum of a patient who had only one persistent left superior vena cava (SVC).

Case Report

A 70-year-old male who had had intermittent brief tachycardia for a year was admitted from our outpatient clinic. His resting echocardiogram revealed atrial fibrillation with a ventricular rate in the normal range. Chest roentgenogram revealed no cardiomegaly. Both the transthoracic wall and transesophageal echocardiography disclosed a cystic mass, 3 cm in diameter, which was attached to the interatrial septum

and bulged into the left atrium. Magnetic resonance imaging showed an isodense cystic mass on T1-weighted image. In addition, a right SVC was absent, while a left SVC was noted, draining via the coronary sinus behind the left atrium into the right atrium (Figure 1). Coronary angiography showed no feeding vessels from coronary arteries.

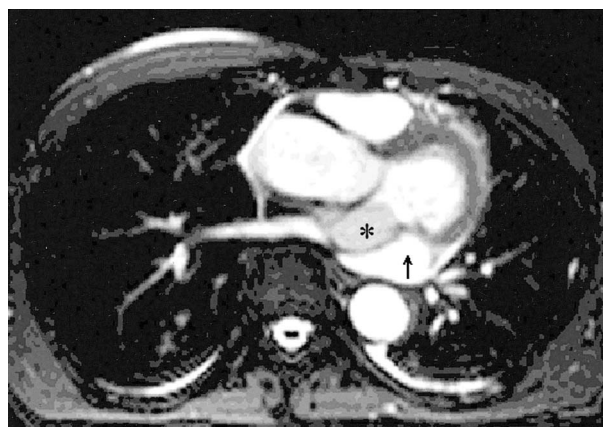


Figure 1. The gadolinium-enhanced T1-weighted magnetic resonance image disclosed a nonenhanced round mass in the interatrial septum (asterisk). A dilated coronary sinus was noted (arrow).

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We performed surgical excision of the cystic mass. After standard median sternotomy, a persistent left SVC was noted on the left lateral side of the pulmonary artery and there was no right SVC. The patient was situs solitus on the atrial level. The heart was arrested under full cardiopulmonary bypass. Both the right and left atria were opened by the “superior-septal” incision (Giraudon incision). The fossa ovalis was absent. The mass was found in the interatrial septum with the intact endocardium overlying it. It bulged into the left atrium and the caudal margin was very close to the conduction system. From the left atrial side, the mass along with its overlying left-atrium endocardium was completely excised out of the interatrial septum (Figure 2A). The septal muscle, which was to the right atrial side of the mass, and the myocardium near the conduction system were preserved. After excision, the left atrial endocardial defect was repaired with autologous pericardium. The left atrial roof was closed directly, while the interatrial septum and the right atrial roof were closed with bovine pericardium. The mass was a soft round cyst, 3 cm in diameter, containing sticky milky white mucus (Figure 2B).

The postoperative recovery of the patient was uneventful. The patient was discharged on the 7th postoperative day. His cardiac rhythm remained as normal sinus during the 1-year follow-up.

Histologic examination revealed that this was a unilocular cyst containing mucus, which had a characteristic pseudostratified ciliated columnar

epithelium and smooth muscular wall (Figure 3). The final pathologic diagnosis of the cystic mass was a bronchogenic cyst.

Discussion

Bronchogenic cysts are classified as a type of congenital foregut cyst. They are usually extrapulmonary and occur mostly along the tracheobronchial tree. As far as we know, there have been only 3 previously reported cases of intracardiac bronchogenic cysts, including 2 in the interatrial septum and 1 in the right ventricle.¹⁻³

The preoperative impression of this case was an atrial myxoma. Other differential diagnoses include lipomatous hypertrophy of the interatrial septum, papillary fibroelastoma, teratoma, primary cardiac lymphoma, etc.⁴⁻⁷ However, the definitive diagnosis of an intracardiac bronchogenic cyst can only be made by pathology. Biopsy of the cyst is a very dangerous procedure because of the risk of rupture of its contents resulting in pulmonary or systemic emboli.¹ Malignancy arising in a bronchogenic cyst has been reported.⁸ Therefore, complete surgical excision of the cyst is indicated for diagnosis and treatment.

The chief complaint of our patient was intermittent brief tachycardia, which indicated an intermittent form of atrial fibrillation.⁹ The atrial septum was partially occupied by the cyst, which impeded the electric conduction from the left to right atrium during normal atrial activation. This gave rise to the

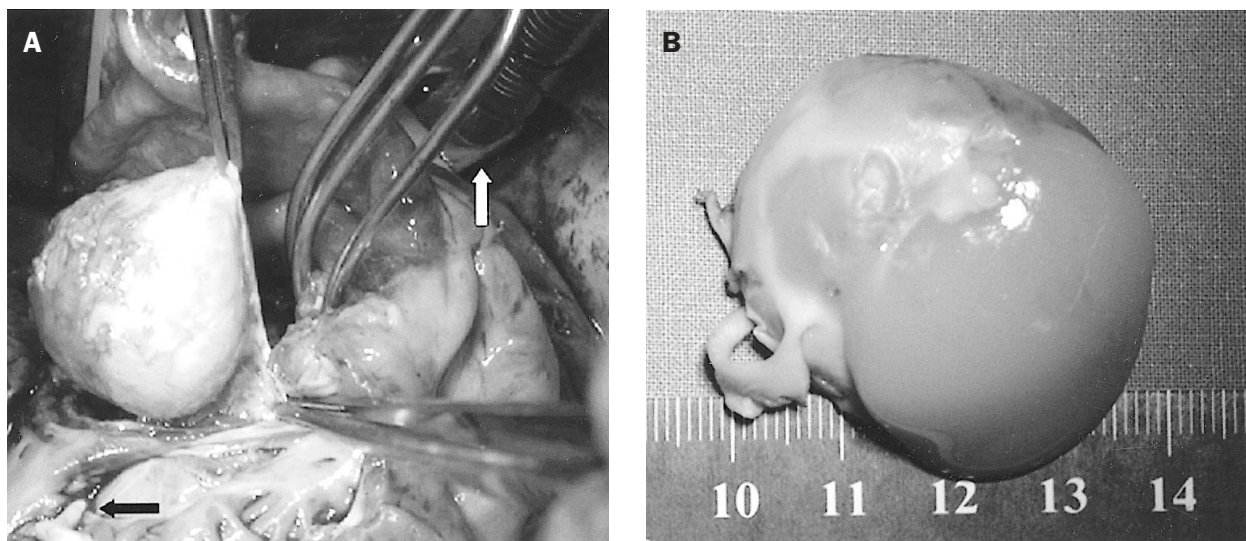


Figure 2. (A) The interatrial cyst was completely excised from the left atrial side. The black arrow shows the Swann-Ganz catheter in the coronary sinus orifice in the right atrium. The white arrow shows the venous cannula in the left superior vena cava. (B) The unilocular cyst was 3 cm in diameter and contained mucus.

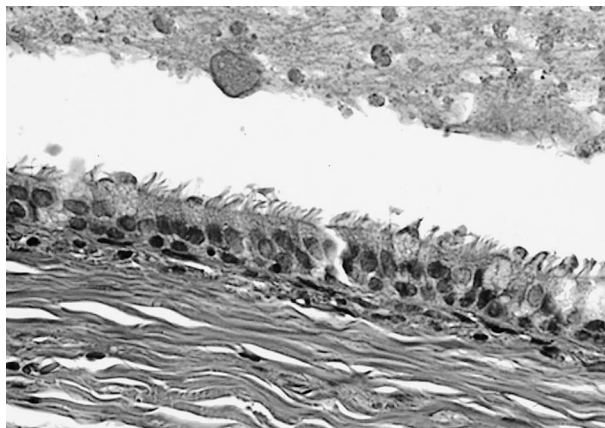


Figure 3. Microscopic examination (hematoxylin and eosin stain, $\times 400$) revealed that the mucus-containing cyst, had a characteristic pseudostratified ciliated columnar epithelium and smooth muscular wall.

opportunity for an automatic focus to initiate macroreentrant circuits and atrial fibrillation accordingly. The opportunity disappeared after removal of the cyst and, hence, the patient returned to normal sinus rhythm postoperatively.

The explanation for the peculiar location of the bronchogenic cyst in this case is as follows. After the 3rd week of embryologic development, there is a ventral diverticulum located in the floor of the foregut. The diverticulum transforms into a tube that becomes the primitive bronchial tree. Abnormal budding of the bronchial tree gives rise to the bronchogenic cyst. The ventral diverticulum derived from the foregut is in close proximity to the heart tube during this early development at the 3rd week, when the heart wall has not developed and the pericardial cavity has not completely surrounded the heart tube. If the cyst arises at the beginning of the 4th week, it may be very close to the primitive heart. As the pericardial cavity grows and gradually surrounds the heart, it includes the cyst in the pericardial cavity. This could explain the existence of the intrapericardial bronchogenic cysts reported

previously.¹⁰ Furthermore, if the cyst is included in the primitive heart during the formation of the heart wall, it becomes an intracardiac bronchogenic cyst. When the cyst is further “deepened” with the infolding of the septum secundum at the 8th week, it becomes the type found in the septum. The muscle of the septum secundum was to the right side of the cyst whose caudal portion occupied the location of the fossa ovalis, which was absent in this case.

This unique case, which is composed of 2 rare congenital anomalies, provides us with an additional differential diagnosis for an intracardiac tumor. Surgical excision of the tumor is necessary for both diagnosis and treatment.

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