Bronchogenic cyst of the atrioventricular septum presenting with ventricular fibrillation

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Introduction
Bronchogenic cysts are one of the types of primary cardiac tumors that can occur. They arise in the interatrial septum, especially near the atrioventricular node, which may cause atrioventricular block owing to its location. The incidence of these cardiac tumors is very low. Previous case reports regarding bronchogenic cysts of the interatrial septum have shown association of bronchogenic cysts with atrioventricular block, but not with the life-threatening ventricular arrhythmias. We report a case with bronchogenic cyst in the atrioventricular nodal region presenting as ventricular fibrillation.

Case report
A 77-year-old woman experienced syncope while walking. She was sent to the emergency department at a local hospital. Twelve-lead electrocardiogram on her arrival showed Wenckebach atrioventricular block and QT-interval prolongation (Figure 1A, QT interval 0.55 seconds) in the absence of electrolyte abnormality. Neither did she have any previous history of QT-interval prolongation to suggest congenital long QT syndrome. After admission, she collapsed with a seizure owing to ventricular fibrillation documented on electrocardiogram monitor (Figure 1B). Atrial flutter was observed after the cardioversion (Figure 1C). An intracardiac mass with 2 cm diameter was identified in the atrioventricular septum by echocardiography, which was further evaluated by cardiac contrast-enhanced computed tomography (Figure 2A). Cardiac magnetic resonance imaging (MRI) performed after she was transferred to our hospital identified a 3-cm-diameter mass in the atrioventricular septal region on T2-weighted images (Figure 2B). Neither fat deposition nor late gadolinium enhancement was identified in the tumor on cardiac MRI images, which led to the diagnosis of a probable cystic tumor. The tumor was successfully resected surgically, but resulted in complete atrioventricular block. During the surgery, the tumor appeared to be compressing the atrioventricular septal region including the atrioventricular node. Electrophysiological testing for the induction of malignant ventricular arrhythmias was performed 3 weeks after the surgery. However, no sustained ventricular tachyarrhythmias were induced even with delivery of triple extrastimuli from the right
ventricular apex and outflow tract during isoproterenol infusion. Based on this result, the patient was only given a dual-chamber pacemaker. During the 6 months of follow-up after discharge, the patient has been free of further cardiac events and symptoms.

Pathology
The resected unilocular cyst contained a grayish white jelly-like fluid (Figure 3A). Histology showed ciliated respiratory epithelium on the inner wall of the cyst (Figure 3B). Smooth muscle cell was also observed within the wall. These histologic findings strongly suggest diagnosis of a bronchogenic cyst.

Discussion
Most such cardiac tumors are benign, although some have caused sudden death owing to hemodynamic and conduction system disturbances. There have been 120 cases of sudden death reported to be associated with primary intracardiac tumors. Bronchogenic cysts are known as a rare tumor located in the atrioventricular septal region that may cause heart block. In fact, there are a few case reports of bronchogenic cyst presenting with heart block. The association of bronchogenic cyst with life-threatening ventricular arrhythmia remains unclear.

This is the first report that has demonstrated the association of bronchogenic cyst with malignant ventricular arrhythmias leading to sudden cardiac death. We hypothesize that ventricular fibrillation in this case occurred as a consequence of QT prolongation owing to bradycardia associated with AV block from compression of the AV node area by the cyst.

The incidence of ventricular fibrillation may have been secondarily affected by compression of the atrioventricular septal region, including the atrioventricular node, owing to the tumor growth because no infiltration of the tumor into the surrounding tissue was observed on histology. In general, bronchogenic cysts may increase in size over time without causing symptoms owing to mucoid production from ciliated epithelial cells. However, our case shows that enlarged bronchogenic cysts may also affect the surrounding tissue. Atrial flutter was recurrently observed in this patient after cardioversion. Compression by the tumor may have facilitated atrial flutter by creating an area of conduction delay in the lower region of the right atrioventricular septum. No observations were made during the surgery that might otherwise explain the atrial flutter. Cardiac MRI is useful for the assessment of cardiac tumors. In this case, cardiac MRI clearly showed an oval tumor in the atrioventricular septal region with T2 high intensity and T1 low intensity and no late gadolinium enhancement, which helped diagnosis of the tumor as a bronchogenic cyst. The presence of structural heart

Figure 2  Contrast-enhanced computed tomography (CT) (A) and T2-weighted cardiac magnetic resonance imaging (MRI) (B) showed a 3-cm mass (red arrows) in the atrioventricular septal region, which was identified as a cystic tumor. LA = left atrium; LV = left ventricle; MV = mitral valve; RA = right atrium; RV = right ventricle; TV = tricuspid valve.
disease, including cardiac tumors located in the septal region, should be assessed by echocardiography and cardiac MRI in patients with marked first- and second-degree atrioventricular block. The early diagnosis of such rare cardiac tumors may provide us with significant information regarding the risk of heart block progression or sudden cardiac death owing to life-threatening ventricular arrhythmias.

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