

Intracardiac Eustachian Valve Cyst in an Adult Detected with Other Cardiac Anomalies: Usefulness of Multidetector CT in Diagnosis

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We present an unusual case of an intracardiac Eustachian valve cyst observed concurrently with atresia of the coronary sinus ostium, a persistent left superior vena cava (LSVC) and a bicuspid aortic valve. There have been several echocardiographic reports of Eustachian valve cysts; however, there is no report of multidetector computed tomography (MDCT) findings related to a Eustachian valve cyst. Recently, we observed a Eustachian valve cyst diagnosed on MDCT showing a hypodense cyst at the characteristic location of the Eustachian valve (the junction of the right atrium and inferior vena cava). MDCT also demonstrated additional cardiovascular anomalies including atresia of the coronary sinus ostium and a persistent LSVC and bicuspid aortic valve.

Index terms: Heart valve; Congenital anomaly; Computed tomography, spiral; Coronary sinus; Eustachian valve cyst

INTRODUCTION

The Eustachian valve, or valve of the inferior vena cava (IVC), is a normal embryologic remnant of the right valve of the sinus venosus (1). The Eustachian valve is thought to regress during childhood, but in some cases, it may persist into adulthood.

Failure of the Eustachian valve to regress may simulate a mass or result in cardiac diseases such as endocarditis, thrombus formation, and vegetations (2, 4). A myxoma arising from the Eustachian valve, although rare, has been

described (3). A solitary cyst attached to the Eustachian valve has also been reported (4).

The anatomic and echocardiographic features of a Eustachian valve cyst have been described. However, to the best of our knowledge, the multidetector CT (MDCT) findings of a Eustachian valve cyst have not been reported to date. MDCT is also useful as a tool to identify complex cardiovascular anomalies at a glance (5).

Herein, we present an adult case of an incidentally detected Eustachian valve cyst at the junction of the right atrium (RA) and IVC that was diagnosed by MDCT. This case was also associated with other rare cardiac anomalies: atresia of the coronary sinus ostium with retrograde drainage of cardiac veins via the LSVC and bicuspid valve with aortic stenosis.

Received August 6, 2011; accepted after revision October 10, 2011.

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CASE REPORT

A 71-year-old man had visited the pulmonary outpatient clinic in our hospital for interstitial lung disease. He was admitted to our hospital due to a sudden worsening of dyspnea and cough. Auscultation of the lungs revealed

inspiratory rates in both lung bases. Laboratory examination demonstrated slightly decreased PaO₂ (80.6 mm Hg) upon arterial blood gas analysis. Otherwise, there were no significant abnormalities in the laboratory data.

A chest radiograph showed cardiomegaly with a cardiothoracic ratio of 0.59 and reticular opacities at the right costophrenic space. Chest CT revealed pulmonary fibrosis consistent with typical interstitial pneumonitis (UIP pattern) showing reticular opacity at the subpleural area in both lungs, especially in the right lower lobe.

Incidentally, a lobulating contoured hypodense mass measuring roughly 1.7 cm was noted at the junction of the RA and IVC. The coronary sinus was markedly dilated, while the coronary sinus ostium was not clearly identified (Fig. 1A). A left superior vena cava (LSVC) was present. Hence, occlusion of the coronary sinus ostium was suspected, and coronary venous flow was presumed to drain through a persistent LSVC. Dense calcification of the aortic valve was also noted. To evaluate the coronary sinus ostium and identify the character of the RA mass, cardiac CT was performed using dual-source CT (Somatom definition; Siemens, Erlangen, Germany; 120 kVp, 320 mAs, 0.33 s/rotation, pitch 0.36, tube current modulation using ECG pulsing, slice thickness of 0.75 mm, 0.5 mm reconstruction increment).

The cardiac CT showed dense calcification in the bicuspid aortic valve with raphe (Fig. 1B). It also showed a lobulating contoured hypodense mass (18 HU) measuring approximately 1.7 cm and without definite enhancement at the junction of the RA and IVC. Differential diagnoses of the mass included, first, myxoma and then thrombus. However, due to their typical locations, a Eustachian valve cyst and Chiari network were also considered.

Continuity of a contrast-enhanced lumen between the coronary sinus (CS) and RA was not noted on cardiac CT; we therefore reported atresia of the CS ostium (Fig. 1C). The dilated coronary sinus continued to the persistent LSVC and then drained into the left innominate vein. It finally drained to the right superior vena cava (RSVC) and into the RA. Volume-rendered CT imaging clearly demonstrated the dilated coronary sinus and cardiac veins (Fig. 1D).

On transesophageal echocardiography, a 2 × 1 cm oval echogenic mass attached to the RA wall was visible (Fig. 1E) and the mass contained an internal anechoic cystic lesion. Moderate to severe aortic stenosis was also diagnosed.

We decided to perform a simultaneous operation for the severe aortic valve stenosis and RA mass excision. During

the operation, when the RA was being opened, a 1.5-cm mass-like sac was found at the junction of the RA and IVC. As soon as the surgeon touched the sac, it burst and dark blood spilled out. The right atrial ostium of the coronary sinus was atretic. The coronary sinus was shown as an aneurysmal dilatation and was connected to the persistent LSVC. Aortic valve replacement and RA mass excision were performed successfully.

On histological examination, the cystic mass was confirmed to be a pseudocyst containing blood (Fig. 1F). The cystic mass was diagnosed as a Eustachian valve cyst, which was supported by the bloody fluid it contained and its typical location at the junction of the RA and IVC.

Finally, we concluded that this patient had a Eustachian valve cyst associated with atresia of the coronary sinus ostium, a persistent LSVC, and a bicuspid aortic valve.

DISCUSSION

The mechanism of Eustachian valve cyst formation is unknown. However, the pathogenesis of a Eustachian valve cyst is presumed to be similar to that of a cardiac blood cyst (4). Cardiac blood cysts are benign cardiovascular tumors that are frequently found in the atrioventricular valves of newborn infants (6). There are two suggested mechanisms underlying cardiac blood cysts. Boyd (7) suggested that cysts are formed during valve development, when blood is pressed and trapped in crevices that are later sealed off, and Sakakibara et al. (8) proposed an injury-related hypothesis known as the "sudden occlusion theory". Similarly, a Eustachian valve cyst might be formed from blood entrapment or from injury-related cyst formation.

The echocardiographic appearance of a Eustachian valve cyst had been reported (4) in addition to a well-defined cystic mass attached to a thickened Eustachian valve in the RA, and it prolapsed intermittently into the IVC on a transesophageal echocardiogram. However, there is, to the best of our knowledge, no report regarding the CT findings of a Eustachian valve cyst.

In our case, the CT showed a well-defined low-density cyst in a typical location of the Eustachian valve. The differential diagnoses of a Eustachian valve cyst include the myxoma, thrombus, and Chiari network. Myxomas typically occur in the left atrium, especially at the interatrial septum. However, 20% of myxomas occur in the RA (9), and there is a case report of a myxoma occurring at the Eustachian valve (10). However, myxomas usually show heterogeneous

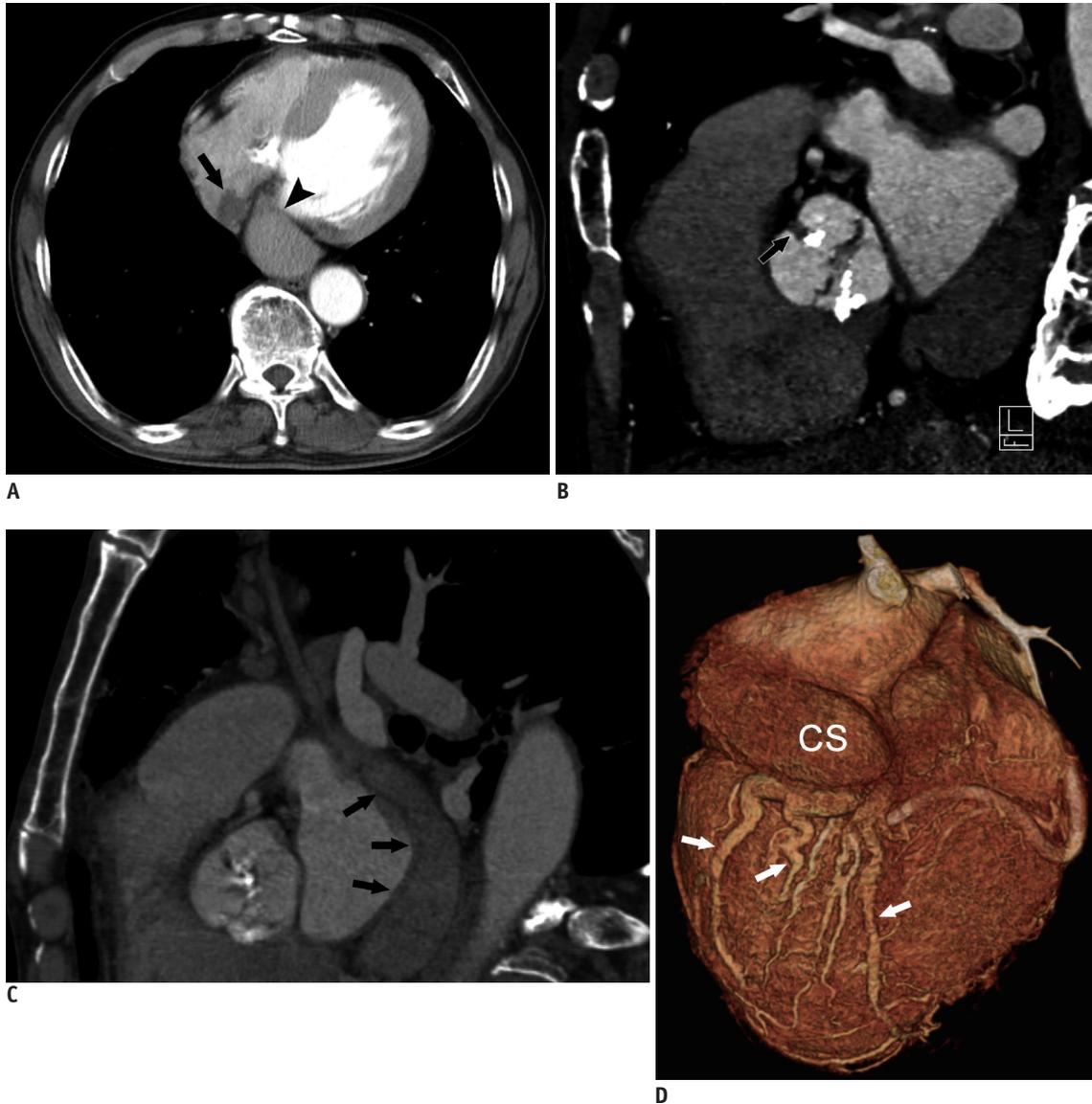


Fig. 1. Intracardiac Eustachian valve cyst in 71-year-old man with other cardiac anomaly including atresia of coronary sinus ostium, persistent left superior vena cava and bicuspid aortic valve.

A. Axial contrast-enhanced chest CT shows 1.7-cm lobulating contoured hypodense mass at junction of right atrium and inferior vena cava (arrow). Note dilated coronary sinus (arrowhead). **B.** Cardiac CT with multiplanar reformatted image shows bicuspid aortic valve with raphe (arrow) and dense calcification, suggesting aortic stenosis. **C.** Sagittal multiplanar reformatted image of cardiac CT shows dilated coronary sinus connected to persistent left superior vena cava (arrows), which drains into left innominate vein (not shown). **D.** Volume-rendered CT image shows dilated coronary sinus (CS) and cardiac veins (arrows).

attenuation due to hemorrhage, necrosis and fibrosis, and some of them are heterogeneously enhanced after contrast infusion on CT (11). Those findings are helpful for the differential diagnosis.

Cardiac thrombus revealed a non-enhancing low-density mass. However, it occurs at chambers of slow flow and in patients with organic heart disease such as atrial fibrillation or infarction (9). The Chiari network is a retinaculum attached to the region of the crista terminalis that extends to the valves of the IVC and coronary sinus or sometimes to

the floor of the RA near the ostium of the coronary sinus. On CT studies, the anomaly often appears as an irregularity or a small soft-tissue density mass along the right atrial wall (12).

Our case was also associated with another rare anomaly: atresia of the coronary sinus ostium with retrograde drainage of the cardiac veins via LSVC. Atresia of the coronary sinus ostium with a persistent LSVC is a rare cardiac anomaly. This condition is usually asymptomatic throughout life, and a majority of cases were reported as

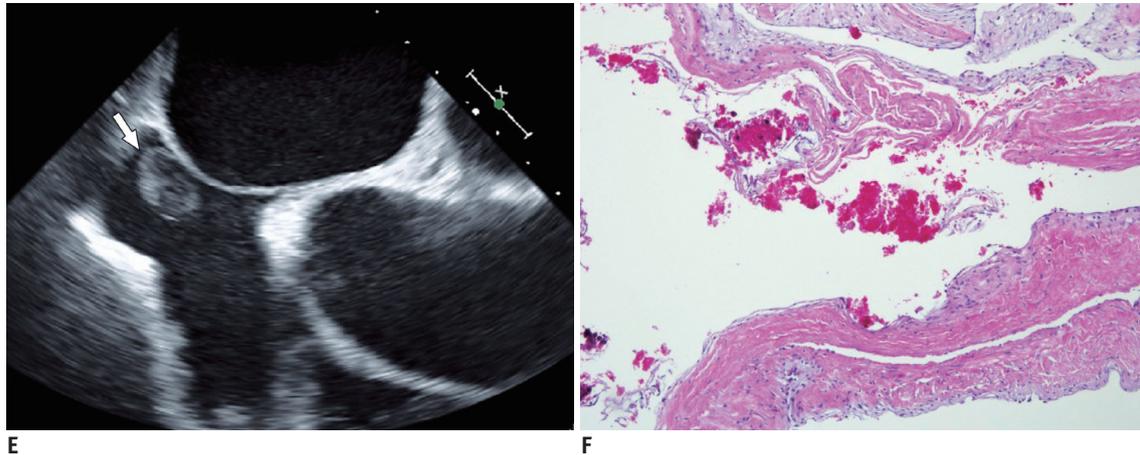


Fig. 1. Intracardiac Eustachian valve cyst in 71-year-old man with other cardiac anomaly including atresia of coronary sinus ostium, persistent left superior vena cava and bicuspid aortic valve.

E. Transesophageal echocardiogram shows 2 x 1-cm oval echogenic mass attached to right atrium wall that contains internal anechoic cystic lesion (arrow). **F.** Pathology shows (H & E stain x 100) pseudocyst induced through fibromyxoid degeneration and flattening of wall with blood.

incidental postmortem findings. In this situation, cardiac venous blood is forced to flow in a retrograde direction via the coronary sinus into the LSVC. The LSVC then drains into the innominate vein and finally into the RA (13). Knowledge of this cardiac anomaly is needed if cardiac surgery is planned.

Atresia of the CS ostium has been reported in association with a number of other cardiac malformations: a univentricular heart, atrial septal defect, ventricular septal defect, transposition of the great arteries, atrioventricular canal, tricuspid atresia, double-outlet right ventricle, partial anomalous pulmonary venous return, pulmonary atresia with intact ventricular septum, and Ebstein's anomaly (14). However, there is no report of an association between atresia of the coronary sinus ostium and a remnant Eustachian valve anomaly or bicuspid aortic valve.

Recently, MDCT has been widely used to diagnose cardiovascular disease and is very useful for demonstrating the complex cardiovascular anomalies at a glance, as in our case (5). In our case, the status of the coronary sinus ostium was initially difficult to evaluate using a non-gated chest CT. Gated cardiac CT allowed for the accurate diagnosis of the coronary sinus atresia, persistent LSVC and the bicuspid aortic valve with calcification, as well as the Eustachian valve cyst. Our case reinforces the usefulness of the gated MDCT for the evaluation of complex cardiovascular anomalies.

In conclusion, we report on a rare case of a Eustachian valve cyst diagnosed by MDCT. MDCT revealed a well-defined, low-density cyst in a typical location of the Eustachian

valve (the junction of the RA and IVC).

Multidetector CT is also useful in identifying other rare anomalies including atresia of the coronary sinus ostium with retrograde drainage of the cardiac veins via the LSVC and bicuspid valve with aortic stenosis.

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