An Incidental Left Atrial Congenital Anomaly

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Clinical History

A 47-year-old man with a history of a prior-excised melanoma presented to his physician with new lower extremity edema. A trans-thoracic cardiac ultrasound was ordered to assess left and right ventricular function, and to exclude a pericardial effusion. His body mass index (BMI) was 38, and thus the study was somewhat limited due to body habitus and poor acoustic windows.

Findings

A trans-thoracic cardiac ultrasound revealed normal biventricular systolic function, without evidence of occult pulmonary hypertension or a pericardial effusion. A poorly visualized “left atrial membrane” was noted, consistent with cor triatriatum sinister (Figure 1). He declined to undergo a transesophageal echocardiogram that was recommended to further delineate this finding, but agreed to undergo a non-invasive cardiac CT evaluation with a second-generation dual source 128-detector row CT scanner. The CT study showed no evidence of a left atrial mass and normal pulmonary vein anatomy. A cor triatriatum sinister was confirmed with unobstructed drainage across a wide fenestration into the left atrium (Figure 2 and Figure 3).

Discussion

Cor triatriatum sinister is a residual membrane in the left atrium, and was initially described by Church in 1868. It is usually diagnosed in infancy and its incidence ranges from 0.1-0.4%, although there are reports of late presentation, as in our patient. The etiology of cor triatriatum sinister is still debated. Theories include incomplete incorporation of the common pulmonary venous trunk into the left atrium, abnormal overgrowth of the septum primum, and entrapment of the left common pulmonary vein by the sinus venosus preventing incorporation with the left atrium. There is usually a communication between the two left atrial chambers due to a fenestration in the membrane, the size of which dictates the clinical syndrome. Large fenestrations rarely lead to symptoms, whereas small fenestrations can cause pulmonary venous hypertension. The differential diagnosis includes a supra-valvular mitral ring. The key distinction of cor triatriatum sinister is its insertion proximal to the left atrial appendage (LAA) as compared to supra-valvular mitral rings, which are located distal to the LAA, and immediately above the mitral valvular plane. The diagnosis can be made by 2D or transesophageal echocardiography, cardiac CT, or cardiac MRI. Second-generation dual-source CT technology was able to offer a high quality diagnostic test in 0.6 seconds, and exposed our patient to a low radiation dose of 4.7 mSv.

REFERENCES

1. Church WS. Congenital malformation of the heart: Abnormal septum in left auricle. Trans Pathol Soc (Lond) 1868; 19:188-90