An Interesting Cause of Eisenmenger's Syndrome

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A 35-year-old female with a history of pulmonary hypertension with secondary polycythemia and congenital asplenia was seen for heart/lung transplant evaluation. Up until 2 months prior, she was able to carry out her activities of daily living without limitation and with very mild dyspnea on exertion. However, her functional status had recently declined significantly, and she had to quit her job. She came in experiencing shortness of breath with minimal exertion, consistent with NYHA III, and was put on 4 L home oxygen via nasal cannula. She reported associated occasional leg swelling and denied any chest pain, syncope, or history of hospitalization.

Electrocardiogram showed a right bundle branch block with a left anterior fascicular block and left ventricular (LV) hypertrophy with T wave inversions in leads II, III, and precordial leads. Echocardiography revealed normal LV size and function (estimated LV ejection fraction 55-59%), enlarged right ventricle (RV) with RV hypertrophy and depressed RV systolic function, single atrial morphology with complete absence of atrial septal tissue (Figure 1), mild mitral and tricuspid regurgitation, and RV systolic pressure of at least 55 above the mean atrial pressure.

Computed tomography angiography (CTA) confirmed cor triloculare biventriculare with partial atrioventricular canal defect. In addition, hepatomegaly, asplenia and normal coronaries, and azygos continuation of the inferior vena cava (also known as absence of the hepatic segment of the inferior vena cava with azygos continuation) were observed.

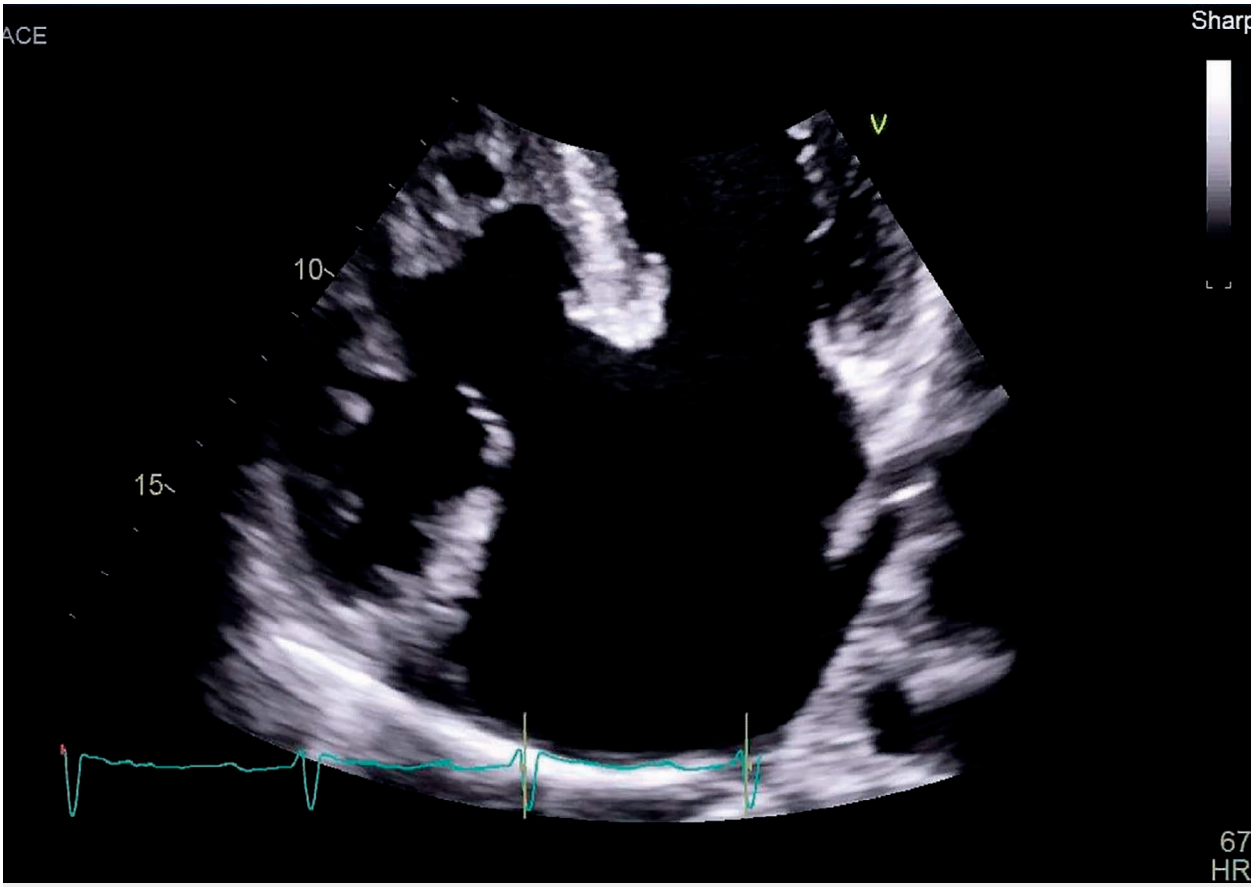

Figure 1.
Apical view in end diastole showing a single atrium.

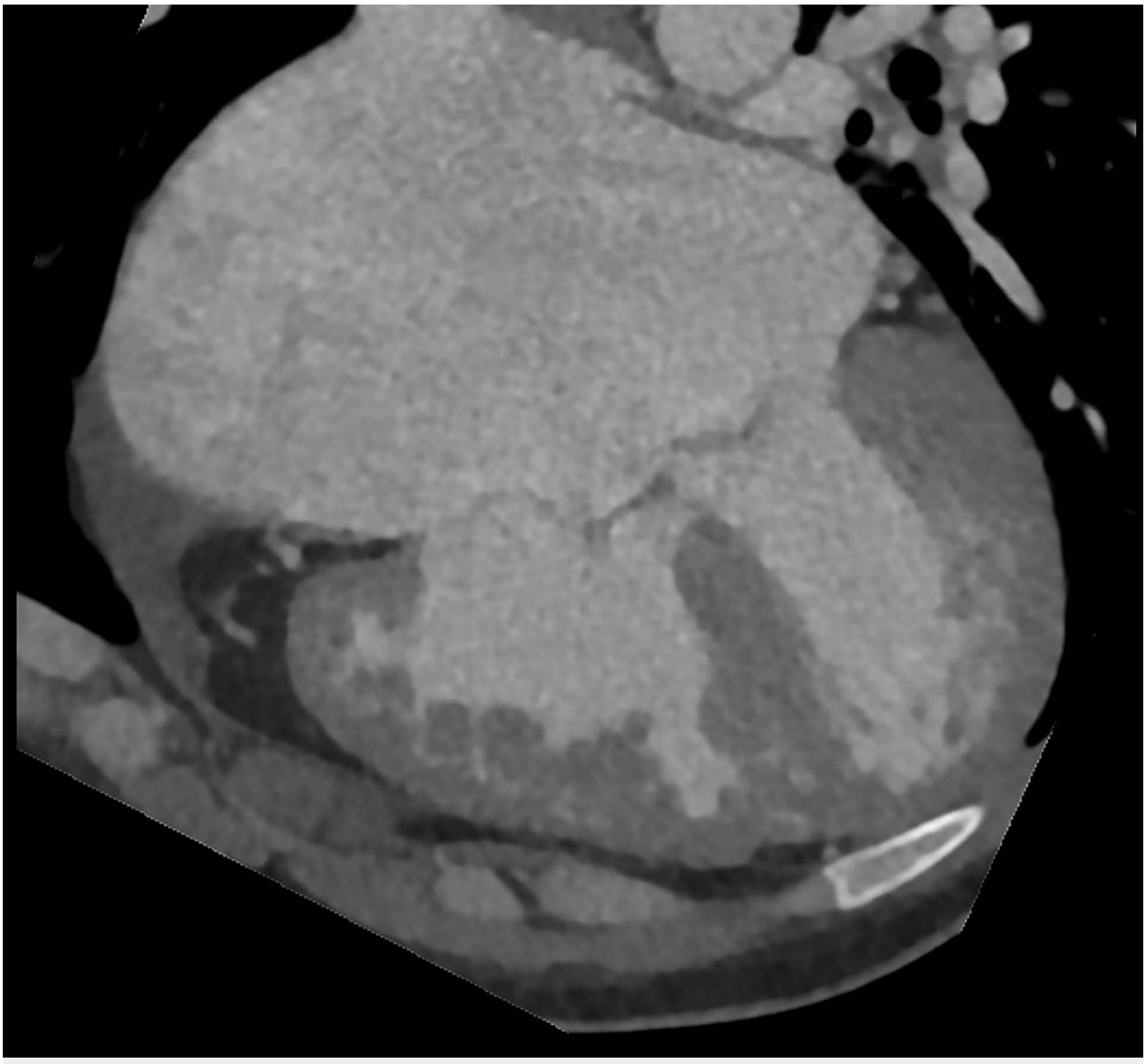

Figure 2.
Three-chamber view revealing the complete absence of the atrial septum.

Cor triloculare, or the three-chambered heart, consists of two atria and one ventricle (cor triloculare biatrium) or, as in our case, two ventricles and one atrium (cor triloculare biventriculare). Both types represent rare congenital anomalies, with the latter being the rarer of the two. Cor triloculare biventriculare results from the lack of septum primum and secundum formation (Figure 2) and presents similarly to a very large atrial septal defect. In the absence of any other cardiac abnormality with Cor triloculare biventriculare, the right atrium and right ventricle dilate and hypertrophy due to the increased amount of blood through the right side of the heart. It was first described by Young et al. in 1907, and few case reports have since been published showing that it may be associated with other congenital abnormalities.[1]

REFERENCES

1. Young AH, Robinson A. Some malformations of the human heart. Med Chron. 1907-1908;47:96-106.