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CASE REPORT

Eustachian valve mimicking atrial thrombosis in a patient with sickle cell disease: A case report

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Abstract

The occurrence of thrombosis in the right heart is an uncommon event, occurring mainly in conditions related to thrombophilia, such as sickle cell disease, or in structural heart lesions or arrhythmias. We describe a case in which an Eustachian valve observed on the transthoracic echocardiogram was confused with an atrial thrombus in a patient with an increased thrombogenic risk. This finding is not clinically important, but may erroneously be interpreted as pathological, and should be differentiated from thrombi and other masses of the right atrium.

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INTRODUCTION

The right atrium often features normal anatomical variations. The Eustachian valve is an embryological remnant of the incomplete absorption of the valve of the inferior vena cava involved in directing oxygen-rich blood from the inferior vena cava through the interatrial septum into the left atrium of the fetal heart. It is a common benign finding in most newborns with unknown prevalence in adults¹.

Viewed on a microscope, the Eustachian valve appears as a fold on the posterior border of the inferior vena cava. In an echocardiogram, it shows as a linear structure stemmed from the junction between the inferior vena cava and the right atrium. It may divide the right atrium into two chambers, producing a defect generally associated with hemodynamic repercussions referred to as cor triatrium². The Eustachian valve is often misinterpreted as disease and has been confused with tumors and thrombi associated with high risk of embolism. Misinterpretation leads physicians to order unnecessary transthoracic echocardiograms and interventions¹.

This paper reports the case of a patient with increased thrombogenic risk whose transthoracic echocardiogram showed a Eustachian valve mistaken for an atrial thrombus. It also presents a brief review on the differential diagnosis of intracardiac masses.

CASE REPORT

A 7-year-old male patient diagnosed at birth with hemoglobin SC disease came to our Hematology service for his annual checkup. Based on his transthoracic echocardiogram, the patient was diagnosed with a thrombus in his right atrium. He was asymptomatic and his physical examination was normal. The patient was admitted to the University Hospital of the Federal University of Juiz de Fora to undergo anticoagulant therapy with heparin and further examination. The rarity of the initial diagnostic hypothesis called for additional investigation by transthoracic echocardiography, which showed he had a Eustachian valve (Figure 1) instead of a thrombus in his right atrium, in addition to a patent foramen ovale. The patient was discharged and has since been followed in our outpatient clinic by a hematologist.

DISCUSSION

The left heart, primarily the left atrium and left atrial appendage, is the most thrombogenic site of the heart³. Although rare, thrombi may also occur in the right atrium, particularly in patients presenting conditions related to thrombophilia, such as sickle cell disease, structural heart disease, or arrhythmia. Sickle cell anemia promotes platelet and plasma coagulation factor activation, thereby creating a state of hypercoagulability and possibly leading to the formation of thrombi via thrombotic microangiopathy. However,

Figure 1. Prominent Eustachian valve in the right atrium (asterisk).



intracardiac involvement is extremely rare, which is why the result of the first echocardiogram of the patient described in this case was challenged⁴.

Echocardiography is a noninvasive imaging method with many uses in clinical practice, including the differentiation of intracardiac masses. The main etiologies of intracardiac masses are cancer, vegetation, and noninfectious masses such as thrombi and embryological remnants⁵.

Heart tumors may originate from the layer of muscle tissue lining the heart or from the pericardium⁵. Primary heart tumors are rare, with myxomas and papillary fibroelastomas topping the list of tumors found in adults and rhabdomyomas in children⁶. Cardiac metastases are more common and originate mainly from lung and esophageal carcinomas and lymphomas⁷. Vegetations are clumps of infected fibrin and clots that may also contain white and red blood cells. They are characteristically seen in patients with endocarditis and may involve natural and prosthetic heart valves, potentially leading to significant morbimortality⁸.

In the case presented, a Eustachian valve - a benign finding without clinical repercussion - mimicked a potentially severe condition known for having a complex and expensive treatment. Reassessment of the patient with an additional echocardiogram corrected the misperception. When performed correctly, echocardiograms allow the differentiation between embryological remnants and signs of disease. Therefore, pediatricians and cardiologists should consider Eustachian valves in their differential diagnoses so that patients are not submitted to unnecessary examination and treatment.

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