

LARGE EUSTACHIAN VALVE - A PUZZLING ECHOCARDIOGRAPHIC DIAGNOSIS AND A DIFFICULT THERAPEUTIC MANAGEMENT

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REZUMAT

Ecocardiografic nu rareori sunt prezente modificări la nivelul atriului drept, date de persistența unor structuri embriologice care nu s-au resorbit. Diagnosticul și conduita terapeutică ulterioară pentru acești pacienți este greu de stabilit. O astfel de structură este valva lui Eustachio, descrisă frecvent și care uneori se însoțește de complicații neașteptate. Raportăm cazul unei paciente cu o masă voluminoasă intraatrială dreaptă, la care, la ecocardiografia transesofagiană s-a ridicat suspiciunea de tromb, dar fără a se exclude cu certitudine o structură embrionară. Pacienta a prezentat asociat regurgitare aortică degenerativă semnificativă, motiv pentru care s-a intervenit chirurgical. Evaluarea intraoperatorie a atriului stâng a arătat o valvă lui Eustachio mare, formațiunea a fost excizată, iar examenul anatomopatologic ulterior a confirmat diagnosticul. Deși prezența valvei lui Eustachio este considerată a fi o situație rară, ea nu trebuie omisă din diagnosticul diferențial al pacientului cu masă intra-atrială dreaptă.

Cuvinte cheie: valva lui Eustachio, ecocardiografie, masă atrială dreaptă.

ABSTRACT

Anomalies of the right atrium, due to incomplete resorption of the embryologic structures, are difficult to diagnose and to establish the future management. One of these structures, the Eustachian valve, is often reported with different unexpected complications. This report presents the case of a patient with a prominent right atrial mass that was suspected at the transesophageal echocardiography to be a thrombus, but the possibility of an embryological remnant was not excluded. The patient had also significant degenerative aortic regurgitation, for which she was operated. The intraoperative right atrial inspection revealed a large Eustachian valve, which was removed. Subsequent pathologic examination confirmed the diagnostic. Although Eustachian valve is usually considered a rare condition, it should not be omitted when evaluating a patient with a right atrial mass.

Key words: eustachian valve, echocardiography, right atrial mass.

INTRODUCTION

Anomalies of the right atrium (RA), due to incomplete resorption of the embryologic right valve of the sinus venosum, the Eustachian valve, are not rare echocardiographic findings. Transthoracic echocardiography (TTE) shows the Eustachian valve in the majority of newborns.¹ The Eustachian valve or valvula venae cavae inferioris has embryologically the role to conduce the oxygenated blood from the

inferior vena cava (IVC) across the patent foramen ovale (PFO) into the systemic circulation.¹

The prevalence of Eustachian valve in adults was not reported in studies until now. Typically, this structure is absent in the adult; when present it is considered to be benign, but an incomplete echocardiographic examination in such patients can yield diagnostic misinterpretations.² When the TTE examination finds a RA mass, the definitive diagnosis is extremely important for therapeutic management and also for patient prognostic. A RA mass is usually a malignant tumor or a thrombus with high risk for pulmonary thrombembolism. Inclusion of Eustachian valve in the differential diagnosis of a RA mass can alleviate concern and spare an unnecessary transesophageal echocardiographic (TEE) examination when the typical TTE characteristics are identified.³

We present a patient with a prominent Eustachian valve associated with atrial septum aneurism, PFO and significant aortic regurgitation.

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CASE REPORT

A 62-years old female was admitted with inspiratory dyspnea and fatigability, symptoms that worsened in the last month. For six years she had been treated with diuretics and angiotensin converting enzyme (ACE) inhibitors for arterial hypertension grade 2. She was also known with degenerative aortic regurgitation grade 2. On physical examination, a systolic murmur grade II and a diastolic murmur grade IV were audible in the 2nd right parasternal space. The chest roentgenogram showed mild enlargement of the heart with a cardiothoracic ratio of 57%. The electrocardiogram showed sinus rhythm at a rate of 60 beats/min and an increase in voltage. Laboratory findings were within normal limits. TTE identified degenerative aortic regurgitation grade III, and a mild dilation of the left ventricle, with type I diastolic dysfunction, normal systolic function, and normokinesis. The right cavities had normal diameters, but in the right atrium appears an echo mobile dense mass. (Fig 1) There was also an atrial septum aneurism. TEE was indicated to visualise the right atrium mass and to complete the evaluation of aortic valve. The RA mass appeared as long, filamentous, approximately 4.5 cm in length, highly mobile and with the insertion at the IVC orifice into the right atrium.(Fig. 2A, B) TEE confirms both the grade III aortic regurgitation through a calcified aortic valve, and the atrial septal aneurism and reveals a PFO.

Complementary investigations were performed: peripheral venous Doppler revealed normal flow, without signs of thrombosis; abdominal echography was also within normal limits.

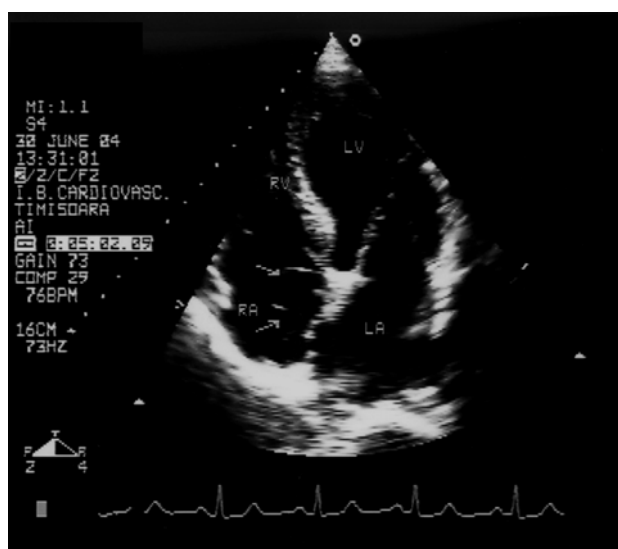


Fig. 1. Transthoracic echocardiography, apical four chambers view; white arrows - right atrium mass; LV = left ventricle, LA = left atrium, RV = right ventricle, RA = right atrium.

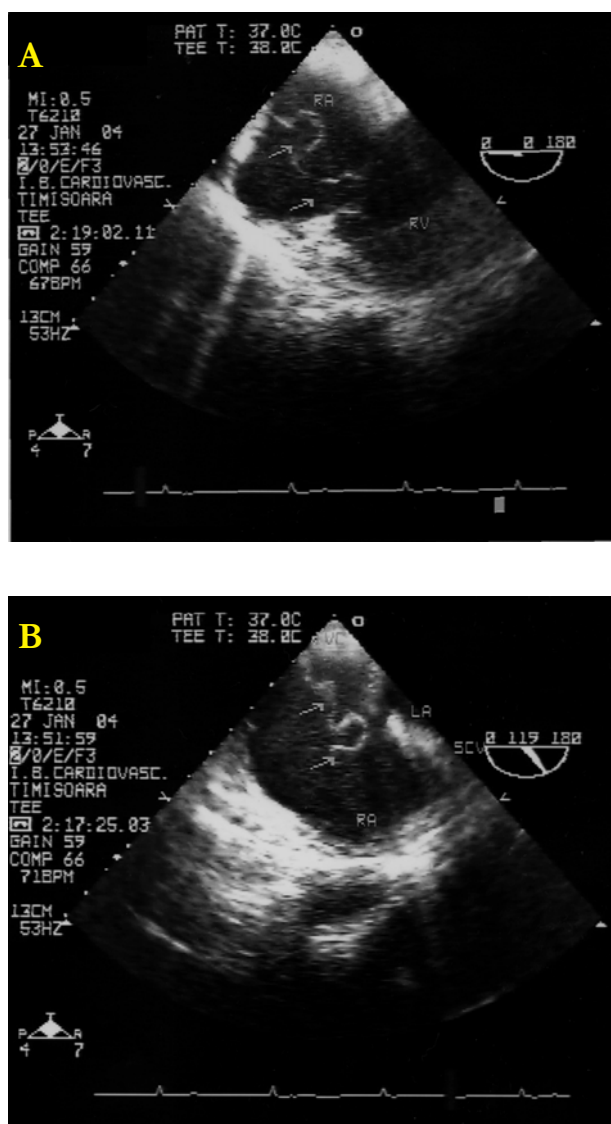


Fig. 2. Transesophageal echocardiography. A. mezosophageal view at 0°, right chambers; B. mezosophageal view at 119°; white arrows - right atrial mass; RA = right atrium, RV = right ventricle, SVC = superior vena cava, IVC = inferior vena cava.

At this moment the surgical intervention was indicated, in order to correct the significant aortic regurgitation and to remove the RA mass. The clinical suspicion for the latter was intracavitary thrombus, but, because there were no favourable factors, the possibility of an embryological remnant, especially an Eustachian valve, was discussed. Coronarangiography performed prior to surgery showed normal coronary arteries.

The opening of the RA revealed a thin, fenestrated, fibrous membrane, at the insertion of the IVC into the RA, criteria which are pathognomonic for Eustachian valve. (Fig. 3) The Eustachian valve was excised as well as the atrial septum aneurism and the foramen ovale closed. A Sorin Bicarbon® (CarboMedics, USA) valve was used for the aortic valve replacement. The patient had an uneventful postoperative course.

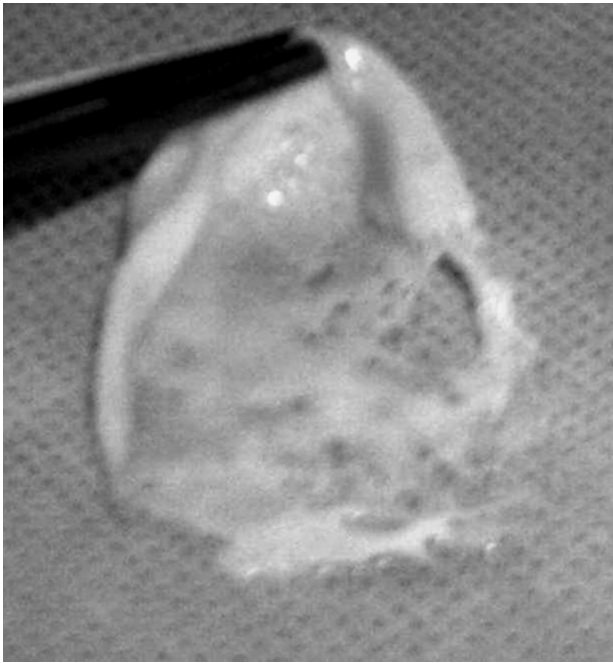


Fig. 3. Prominent Eustachian valve, surgically removed.

DISCUSSION

Several acquired or congenital structures can make the differential diagnostic of an echo dense RA mass, including: thrombus, vegetation, tumour, embryonic remnants, Chiari network and Eustachian valve.⁴ In the present case the differential diagnosis for the echo dense mass included the entire list, and the preoperative investigations could not make a certain diagnostic. This problem was easily resolved in the operation that was indicated for significant aortic regurgitation. During surgery, the diagnostic for RA mass was clear for Eustachian valve because of its characteristics: a laminar dense structure that originates at the orifice of the IVC, crosses the floor of the RA and inserts into the septum primum, corresponding to the descriptions in the literature.^{4,5} The diagnosis was confirmed by histological examination, which showed fibrous conjunctive tissue.

But what would have happened if the patient had not had a significant aortic regurgitation? If we had considered the RA mass to be an embryologic remnant then the surgery would not have been indicated. The echocardiographic signs - a filamentous mass with the origin at the orifice of IVC - suggested an Eustachian valve. The association between Eustachian valve and PFO reported in the literature was another reason for supporting this diagnosis.¹ But the dimensions of the RA structure raised the suspicion of an thrombus migrated from the veins of the lower extremities or an Eustachian valve thrombus. In both cases the risk for pulmonary thromboembolism is high. The last example, a thrombus in the right atrium attached to

the Eustachian valve, although an uncommon site for thrombus formation, it is not rarely described in the literature.⁶⁻⁸ It is also known that Eustachian valve can indirectly predispose to paradoxical embolism, by directing the blood from the inferior cava to the interatrial septum, and through an PFO in the left atrium and then in the systemic circulation.² Moreover, a persisting Eustachian valve may prevent spontaneous closure of PFO after birth.⁶

Although the Eustachian valve is an uncommon site for a vegetation to be attached, there are reports that reveal the possibility of infection with vegetations here, vegetation that can also embolise.^{9,10} The diagnosis of infective endocarditis can be missed because a systematic approach of vegetation is not routinely performed to search for vegetations on this structure. The accidental echocardiographic finding of an Eustachian valve is not an indication for infective endocarditis prevention.

Can all these facts support the indication for surgical removal of the Eustachian valve? Probably for a large structure, but nothing is standardized. The decision is a result of a combined cardiac and surgical exam.

In fact, there are reports suggesting that the Eustachian valve can persist in adults and cause or simulate cardiac problems,¹¹⁻¹⁸ but there are no systematic data on the prevalence or clinical importance of Eustachian valve.

In these cases data from TTE and TEE views may be sub-optimal for examining the entire right atrium and the posterior structures of the heart that need to be investigated in detail for a complete diagnosis. During the examination potentially false images can appear; that need to be known for correct image interpretation. Echocardiographic three-dimensional reconstructions of RA masses can provide information to clarify the diagnostic of embryonic remnants or other pathologic findings in order to avoid unnecessary surgical interventions.^{19,20}

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