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Short Communication

Giant Right Atrial Aneurysm: A Rare Congenital Heart Disease

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ABSTRACT

Congenital right atrial aneurysm is a rare congenital heart disease defined as a dilation of the right atrium in the absence of an underlying cause. The clinical presentation varies; most patients are asymptomatic, while others may experience arrhythmias, dyspnea or intracavitary thrombi. Diagnosis is made by echocardiography. Medical and surgical management remain debated. Surgical resection is indicated for symptomatic patients.

Keywords: Right atrial aneurysm; Congenital heart disease; Echocardiogram

Abbreviations: AR: Atrial Right; RV: Right Ventricle; RAA: Right Atrial Aneurysm; LV: Left Ventricle; CMRI: Cardiac Magnetic Resonance; CT: Cardiac Tomography

INTRODUCTION

Right atrial aneurysm is an uncommon condition, with few cases described in the literature [1-3]. It is characterized by an isolated enlargement of the right atrium in the absence of additional cardiac lesions causing right atrium dilatation. These aneurysms can be congenital or acquired and they could affect both left and right atrial appendage. In congenital cases, the cause is a muscular wall dysplasia with a lack of pectinate muscles of the right atrium. Only there is a remaining part of a muscular rim near the crista terminalis and the superior vena cava [2-4]. Furthermore, endocardial fibrosis and lipomatous degeneration without inflammatory reaction are observed. Associated congenital heart disease have been described such as atrial septal defect, persistent left superior vena cava and anomalous pulmonary drainage.

DESCRIPTION

Clinical presentation is variable. Patients with this condition may be asymptomatic initially, manifesting mainly in adulthood [4]. Most of cases are incidentally diagnosed. Common clinical manifestations include dyspnea, palpitations, supraventricular arrhythmias (atrial fibrillation or flutter) [5]. Symptoms of right heart failure are less frequent [6]. Complications can include thrombosis, pulmonary embolism, atrial rupture and sudden death. It is believed that the cause of the arrhythmias is atrial dilatation and structural disorientation of the myocardial fibers and conduction system that cause ectopic foci or reentrant circuit [7,8]. Two reported pediatric cases described symptomatic airway obstruction and was reversed after aneurysm resection [1].

Before performing trans catheter VSD closure, it's important to consider the potential risk of atrioventricular block. TEE is a useful tool for imaging the VSD anatomy and its relationship with surrounding structures. During the procedure, there is a possibility of conduction system injury from mechanical trauma or compression caused by the delivery system or device, which could result in acute intra procedural atrioventricular block. Patients and their family should be made aware of the symptoms and seek immediate medical attention if necessary. Follow-up care is also important for patients with implanted devices both immediately and in the long-term. Future device modification (softer and less traumatic occluders) could probably help to avoid the occurrence of cAVB.

Diagnosis is often made only by transthoracic echocardiography, mainly in pediatric patients. It assesses the degree of RA dilation, the size of the right ventricle, which may appear small due to external compression and the presence or absence of thrombi. Other structural defects and causes of RA dilation must be ruled out like Ebstein anomaly. Evaluation should be complemented with other imaging methods such as multidetector computed tomography or magnetic resonance imaging to assess anatomical relationships and measure volumes. Fetal echocardiography often provides adequate imaging for prenatal diagnosis.

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The management of RA aneurysms is still debated. Conservative management with anticoagulation or aspirin and antiarrhythmic is an option for asymptomatic patients or those with mild to moderate dilation. Some groups prefer low-dose aspirin for thromboprophylaxis due to the low risk of adverse effects. Regular follow-ups are recommended, including CT/CMR evaluations every 2 years. Pediatric indications for surgery include symptomatic patients, atrial arrhythmias, intracavitary thrombus, major atrial dilatation and severe tricuspid regurgitation due annular compression or those with right ventricular compression or adjacent structures despite symptoms. Some groups advocate for early surgical intervention, even in asymptomatic patients, due to the high potential for severe complications and to prevent further RA dilation [9]. The surgery is low-risk with excellent mid-term results. Most reports used extracorporeal circulation and the techniques involved aneurysm resection and direct suturing or using a pericardial patch to close the atrial wall defect. Injury of the right coronary must be avoided because it could be protruding onto aneurysmal sac. Postoperatively, recurrence of arrhythmias or embolic phenomena is generally not observed.

CONCLUSION

Right atrial aneurysms are a rare congenital disease that can lead to potentially fatal events. Clinical and surgical management is still debated. Patients with large or symptomatic aneurysms should undergo surgical resection. For asymptomatic patients or those with smaller aneurysms, conservative management with anticoagulation or aspirin and regular monitoring could be considered.

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