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Twisted Heart with Atrial Septal Defect: An Interventionalist Nightmare: Cases Reviewing an Absent Etiology

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Abstract

Congenital absence of the pericardium is a well described but rare malformation which occur either as complete absence of the entire pericardium or as a partial left or right portion of pericardium. Cardiac anomalies are common with congenital absence of pericardium which includes Atrial septal defect, Patent ductus arteriosus, Mitral valve disease, Tetralogy of Fallot and Sinus venosus defect. We report two cases of retrospective diagnosis of congenital absence of pericardium after Atrial septal defect device closure which masqueraded as device embolization.

Introduction

Congenital pericardial defects are rare with reported incidence of <1 in 10,000 and underestimated true prevalence due to asymptomatic and incidental diagnosis [1]. The most common variant is partial absence of pericardium mainly left sided defects (70%) followed by right sided defects (17%) and least common complete bilateral absence of pericardium (9%) [2]. Majority of the patients are asymptomatic and are diagnosed incidentally on chest radiographs or while undergoing cardiac surgery. Symptoms are often vague with chest pain, shortness of breath or palpitations and may also arise from associated cardiac anomalies. Atrial Septal Defects (ASD) are common cardiac anomaly found in congenital absence of pericardium. Transcatheter device closure of the atrial septal defect is a feasible option but may pose a dilemma regarding the position of the device post procedure due to leftward and posterior shift of the cardiac silhouette in congenital absence of the pericardium. We report two cases of ASD device closure which presented as device embolization and on further evaluation both the cases were found to have congenital absence of the pericardium.

Case Series

Case 1

A 4 years female came to our center with chief complaints of difficulty in breathing in the past 3 years of age. She was first in birth order born out of nonconsanguineous marriage at term with normal vaginal delivery and had no postnatal complications. She had history of recurrent episodes of lower respiratory tract infections requiring hospitalizations for which cardiac consultation was advised. On examination, the patient was hemodynamically stable with systemic pressures of 111/64 mmHg, respiratory rate of 30 beats per minute and SpO_2 of 100% on room air. On cardiovascular examination first heart sound was normal with wide and fixed splitting of second heart sound and soft mid diastolic murmur at left lower parasternal area. Chest radiograph revealed dilated right atrium and right ventricle with increased pulmonary blood flow. A 12 lead ECG showed sinus rhythm with evidence of right atrium and right ventricular volume and pressure overload.

Echocardiography demonstrated large malaligned ostium secundum ASD measuring 16 mm with adequate rims, enlarged Eustachian valve, dilated right atrium and right ventricle and ventricular function was normal. Transcatheter closure of the ASD was done with 18 mm Amplatzer Septal Occluder without any complications. Post-procedure TEE showed device in position with no residual shunt. On the next day transthoracic echocardiography showed ASD device to be embolized in Left atrium with loss of atrioventricular rim. The child was taken for surgical ASD device retrieval and defect closure. During surgery it was found that there was absence of left posterior and mediastinal pericardium with ASD device attached to the septum. On retrospective analysis of chest X-ray revealed leftward and posterior rotation of cardiac silhouette with straightening and

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elongated left heart border and the space between aorta and the main pulmonary artery appeared widened 'Snoopy Sign'. It was concluded that false presentation of device embolization was due to congenital absence of left posterior and mediastinal pericardium.

Case 2

An 8 years male child, second in birth order, full-term with no postnatal complications was coincidentally found to have ostium secundum atrial septal defect while evaluating for increased precordial activity. On cardiac examination first heart sound was normal, second heart sound was wide and fixed split and a short ejection systolic murmur was heard at left lower sternal border. Chest X-ray demonstrated cardiomegaly with prominent pulmonary arteries and increased vascularity. Transthoracic echocardiography revealed moderate sized ostium secundum ASD with adequate rims and dilated right atrium and right ventricle. Child was taken for transcatheter closure of ASD which was done with 16 mm Amplatzer Septal Occluder. There were no procedural complications and child was discharged on oral antiplatelet. However, on follow-up child had complained of chest discomfort and on evaluation with ECG was normal and Echo showed stable device position with no residual shunt and no evidence of erosion/effusion. Child was kept on follow-up. On 10 months of follow-up in view of persistent chest discomfort CT pulmonary angiography with coronary evaluation was done which showed levorotation of heart with air interposition between PA and aorta and below the heart with prominent left atrial and normal device position and normal coronaries giving the diagnosis of congenital absence of the pericardium. Retrospective evaluation of the chest X-ray showed levorotation of the heart with obscured right heart border and poor visualization of device due to its superimposition on spine and prominent LA with air seen in aortopulmonary window area in left border suggestive of absent pericardium.

Discussion

Congenital Absence of Pericardium (CAP) is exceedingly rare cardiac anomaly which occurs due to failure of pleuropericardial membranes to fuse completely on one or both sides. Ellis classified pericardial agenesis into various subtypes namely complete or partial left sided defect, complete or partial right sided defects and diaphragmatic pericardium defects [3]. Left sided defects are the most common accounting for 70% of all the defects. Diagnosis of CAP is challenging due to asymptomatic presentation or nonspecific symptoms like atypical chest pain, palpitations, dizziness, dyspnea and trepopnea [4]. Majority of the cases are diagnosed incidentally on chest X-ray due to levoposition of the heart and during surgery for other associated cardiac anomalies. About 30% of the CAP have associated cardiac anomalies namely atrial septal defects, patent ductus arteriosus, mitral valve disease, Tetralogy of Fallot and sinus venosus defect. Chest X-ray shows leftward shifted cardiac silhouette, elongated left heart border, cardiomegaly, radiolucency between aorta and pulmonary artery due to lung tissue also known as Snoopy heart sign [5]. ECG demonstrates right axis deviation, right bundle branch block in V1 and poor R wave progression. Echocardiography may show right ventricular dilatation, hypermobility of heart, posterior orientation of the apex, elongated atria with widened ventricles give tear drop appearance of the heart. Echocardiography may not be diagnostic but the above finding should make clinicians aware about congenital absence of pericardium. Meanwhile cardiac MRI remains gold standard for diagnosis of CAP with high sensitivity demonstrating the absence of preaortic pericardial recess which is present in normal hearts. Despite these advancing modalities majority of the cases often goes undiagnosed and has been diagnosed incidentally during surgery or CT angiography which was seen in our cases too. Though complete defect of the pericardium have excellent prognosis but partial absence of pericardium can lead to tricuspid regurgitation by traction of chordal structures, fatal myocardial strangulation, ischemia and sudden death requiring surgical intervention [6,7]. There are literatures which mentions about the rarity of this congenital anomaly and challenges during diagnosis of this entity [2-5,8]. Our cases were much different from the described case reports because none of the literatures mentions about the ASD device orientation which masqueraded as embolization due to levorotation of the heart. Surgical intervention could have been avoided in our first case as false presentation of device embolization was due to levorotation of the heart seen in congenital absence of pericardium. Similarly in the second case persistent chest discomfort and device orientation in chest X-ray and echocardiography could unnecessarily lead to surgical intervention in the child for device retrieval and defect closure if diagnosis of CAP was not made. In both the cases suspicion of device embolization led to further workup and diagnosis of congenital absence of pericardium was made which would have been missed otherwise.

Conclusion

This case emphasizes that clinicians should be aware that ASD device embolization can sometimes be a false presentation due to levorotation of the heart. These features may clinch diagnosis for congenital absence of pericardium and clinicians should retrospectively analyze imaging modalities for presence of congenital absence of pericardium.

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