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Congenital Rudimentary Posterior Mitral Valve Leaflet Diagnosed in Adulthood Associated with Giant Left Atrium and Complete Absence of Pericardium.

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Abstract

Hypoplasia of the posterior mitral valve leaflet (PMVL) is a very rare mitral valve anomaly, and late presentation in adulthood is very rare because of the high morbidity and mortality generated in childhood. Its association with giant left atrium and congenital absence of the pericardium (CAP) is an extremely rare and not reported in literatures till now.

CAP is a very rare anomaly. The majority of cases are asymptomatic, but some may present with nonspecific symptoms.

We report a 53 year old male, a known case of severe mitral regurge (MR) diagnosed 20 years ago, he complained of breathlessness on mild exertion, Echocardiography revealed mitral valve disease which was indicated for mitral valve replacement (MVR) but he refused the surgery at that time.

Keywords: Hypoplasia, congenital anomalies, mitral regurgitation, mitral valve.

Introduction

Hypoplasia of the PMVL is a rare congenital heart disease and is usually presented in infancy and childhood with severe MR, either in isolation or associated with other cardiac lesions. The absence of PMVL is usually symptomatic, due to severe MR and coexisting abnormalities such as intracardiac shunt (1).

CAP is one of the rarest cardiac congenital anomalies. It can occur as a complete absence of the entire pericardium, absence of the right or left portion of the pericardium or a partial, foramen-like defect of the right or left pericardium, while the majority of cases

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are clinically silent, multiple reports associate CAP with symptomatic presentation. The most critical complication of CAP is sudden cardiac death (SCD) due to cardiac strangulation across a partial defect of the pericardium. Given its rare occurrence, most clinicians and imaging specialists will have little experience with this condition and may fail to recognize it on studies (2).

Huge left atrium (LA) is mostly occurred in patients with rheumatic mitral valve disease with severe MR and or mixed stenotic with regurgitation. Many compressive symptoms may occur such as esophageal compression causing dysphagia and tracheal compression causing hoarseness of voice and shortness of breath (3).

Case presentation

A 53-year-old male was admitted with dyspnea on mild exertion (NYHA class III) associated with palpitation with progressive course over the last 3 months. He had been diagnosed with mitral valve disease 20 years ago by echocardiography, which revealed rudimentary PMVL without any attachment of chordae tendinae or papillary muscles, flail anterior mitral valve leaflet (AMVL) and torrential mitral valve regurgitation with mitral valvular area of 4.1 cm2, hugely dilated left atrium 14 x 11 cm, severely impaired right ventricle function (TAPSE 0.9 CM), mild tricuspid regurge and severe pulmonary hypertension (70 mmHg).

Physical examination revealed pulse rate 86/min, atrial fibrillation (AF), blood pressure 115/60 mmHg, his neck veins were distended, and grade III parasternal heave. There was soft and variable S1, wide split S2, pan systolic murmur at apex. Electrocardiogram showed AF with controlled ventricular rate. His chest radiograph revealed massive cardiomegaly with cardio-thoracic ratio of 0.80, left ventricle type of apex, dilated left atrium.

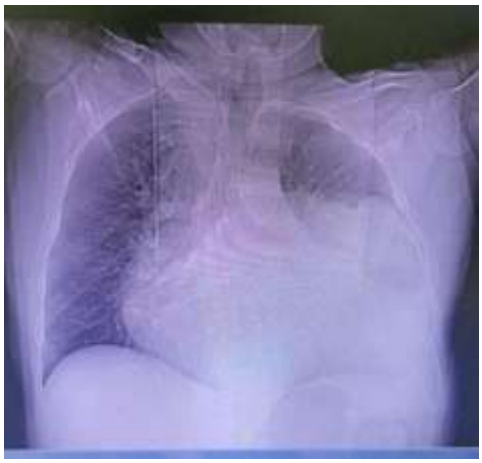

Figure 1: pre operative chest X ray shows hugely dilated left atrium

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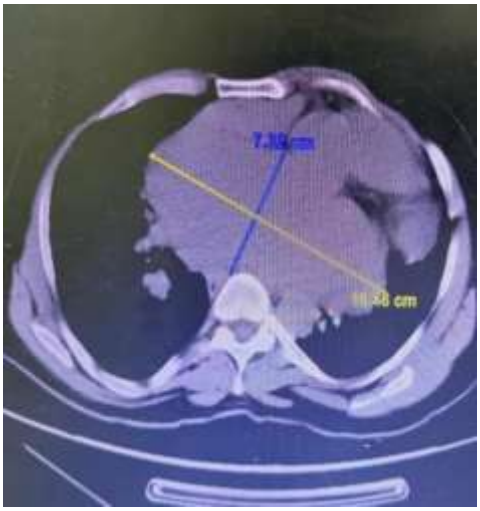

Figure 2: pre-operative CT aortography shows hugely dilated left atrium

Figure 3: pre-operative echocardiography shows hugely dilated left atrium.

CT aortography was done revealing hugely dilated left atrium with dimensions 16.4 x 7.3 x 10.3, volume 1260 ml with significant compression of the right atrium (RA) and right ventricle (RV). The patient was treated with diuretics, oral anticoagulation, and βblocker and was planned for surgical MVR and LA size reduction. Coronary angiography was done and revealed normal coronary arteries. The patient was cleared from dental Septic foci.

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Surgical procedure

The surgery was done under general anesthesia, right central line, arterial line and urinary catheter were inserted (patient has hypospadias). Classic median sternotomy was done revealing complete absence of the pericardium and left pleura, aorto-bicaval cannulation. Using ante and retro grade cardioplegia and snaring of both SVC and IVC were done.

Standard left atriotomy was done, excision of anterior mitral valve leaflet with preservation of the rudimentary posterior leaflet, the mitral valve was replaced by metallic On-X metallic 25-33 valve and reduction of the left atrial wall was done.

Intravenous infusion of Milrinone was loaded and maintained. Weaning from cardiopulmonary bypass was smooth, hemostasis and closure of the wound in layers were done. Post-operative course was uneventual and the patient was discharged home in day 7 post surgery, the patient was followed up for 3 months post-operative in out-patient clinic with marked improvement of his symptoms.

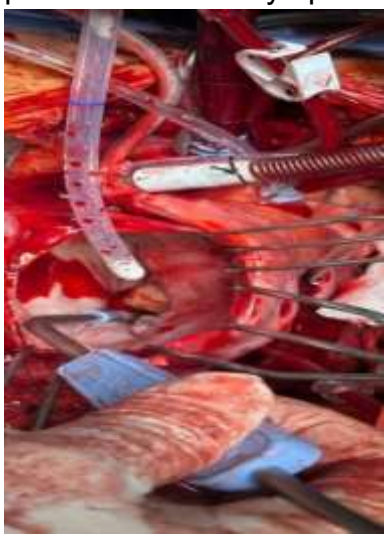

Figure 5: surgical view of the huge left atrium with rudimentary posterior mitral valve leaflet

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Discussion

Congenital mitral valve malformations with late adulthood presentation associated with giant left atrium and complete absence of pericardium are extremely rare combination.

Hypoplasia of the PMVL has been reported and a few cases of absent PMVL have been described. Hypoplastic PMVL is one of the rarest MV anomalies, with an incidence of approximately 1:8, 800 patients according to a prospective study involving 26,484 participants in a routine echocardiographic examination. This pathology has been most frequently observed in infants and children with symptomatic MR, either in isolation or in association with other cardiac lesions, with a tendency to be incompatible with life in many cases. However, some cases have been described in adults, with ages ranging between 17 and 76 years (4).

The antero-posterior diameter of left atrium greater than 80 mm on transthoracic echocardiography is considered diagnostic of giant left atrium. Rheumatic heart disease with MR and or combined lesion represents the main cause of left atrium dilatation. Giant left atrium can cause intracardiac or extracardiac compression manifestations such as shortness of breath, dysphagia, hoarseness of voice, palpitations, chest pain, swelling of the body and thromboembolic events (5).

Additional imaging modalities should be considered to precisely assess the size and its relationship with surrounding structures. The largest left atrial diameter measured in the literature was 20 x 22 cm in a patient with a prosthetic mitral valve (6).

Management of severe MR with giant left atrium is surgery to correct the mitral valve abnormalities, treat compression manifestations, prevent thromboembolism, and revert atrial fibrillation to normal sinus rhythm. Mitral valve surgery with or without left atrium volume reduction is indicated in giant left atrium. The main indication for volume reduction is the presence of intracardiac or extracardiac compressive symptoms (7).

In our case report combined pathology of complete absence of pericardium and severe congenital MR with hugely dilated left atrium with late manifestation lead us to ask question: Can complete absent pericardium facilitate the development of giant left atrium as a result of associated mitral pathology?

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consents

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Conflicts of interest

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There are no conflicts of interest.

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