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Case report

Submitral aneurysm of the left ventricle

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KEYWORDS

Aneurysm
Cardiac
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ABSTRACT

Submitral aneurysm is a rare cardiac pathology of uncertain origin with varied clinical manifestations. Recent studies have revealed a congenital basis of this pathology, although genetic link has been suspected because of the racial predilection. The other suggested aetiologies are infection and inflammation. The case reported here is that of a young female with a large submitral aneurysm presenting in a state of cardiogenic shock. In addition, the presence of raised inflammatory parameters indicates that the cause of origin of this aneurysm is related to inflammation.

Introduction

Submitral aneurysm of the left ventricle (LV) though relatively unknown is a widely recognised cardiac pathology of varied aetiology. Reported for the first time in Nigeria and other African nations, this disease is more prevalent among the black Africans.[1] While this racial predilection is still prevalent, cases have been described in patients of other races from different parts of the world including India. Patients with submitral aneurysm exhibit varied clinical manifestations,[2–4] and are reported to have poor surgical outcome. We report a young female with a large submitral aneurysm of the LV who presented in cardiogenic shock.

Case report

A 27-year-old female presented with a history of severe retrosternal chest pain with progressive breathlessness of 2-week duration and a low-grade intermittent fever-associated with these symptoms. She was referred with the diagnosis of pericardial effusion causing cardiac tamponade, and already on anti-tubercular drugs. On general examination, heart rate was 110/min, blood pressure 86/60 mmHg, with cold and clammy extremities. Cardiovascular examination revealed soft heart sounds without any audible added sounds. She had low haemoglobin count (8.6 g/dL), high total counts (17,600/mm^3 of blood) with polymorphonuclear leucocytosis, and positive C-reactive protein. Resting electrocardiogram (ECG) showed sinus rhythm with low-voltage complexes, and chest radiograph revealed grossly enlarged cardiac silhouette with a cardiothoracic ratio of 18/25.5 cm, straight left cardiac border and normal pulmonary vasculature. Transthoracic echocardiogram (TTE) revealed moderate pericardial effusion without cardiac tamponade with preserved LV function. Of significance was the presence of an aneurysmal dilatation of the LV in its posterior area adjacent to the mitral valve with an ostial communication between the aneurysm and this cardiac chamber. Transoesophageal echocardiography (TEE) confirmed the findings (Figure 1A). Computed tomography (CT)-angiogram showed a large aneurysm of the LV arising from the posterolateral wall with a wide neck situated close to the mitral valve. There was moderate pericardial and right pleural effusion with a mild degree of mediastinal lymphadenopathy. Coronary arteries were reported as normal. Since, cardiac catheterisation could not be performed due to rapid deterioration of the patient's condition, surgical treatment was opted for.

Surgery was performed under cardiopulmonary bypass. During surgery, the inspection after median sternotomy revealed inflamed pericardium with thick exudates. A large aneurysm was visible involving the posterolateral wall of the LV, measuring 10 × 8 cm with part of the posterior wall adherent to the pericardium (Figure 2A). The neck of the aneurysm was wide measuring 5.5 cm with distinctly visible chordae

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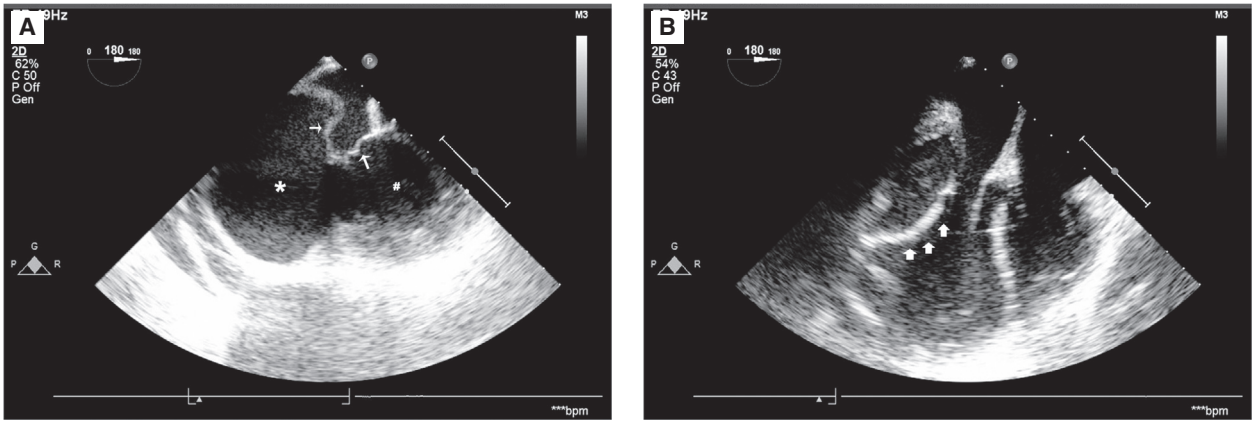

Figure 1 (A) Transoesophageal echocardiogram showing a large submitral aneurysm. (*) indicates the aneurysm; (#) indicates the left ventricle; small arrows indicate the mitral valve. (B) Transoesophageal echocardiogram immediately after surgery showing the Dacron patch (thick arrow) used during surgery to repair the aneurysm.

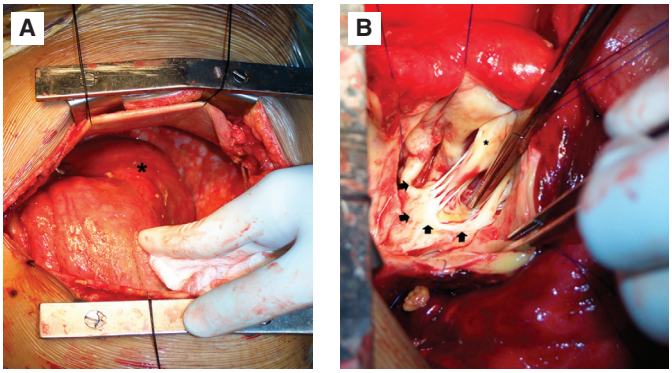

Figure 2 (A) Intra-operative picture image of the submitral aneurysm (black star) after opening the pericardium. (B) Intra-operative picture image after exposing the neck of the aneurysm (small black arrows), showing the papillary muscle with its chordae within the aneurysm.

and papillary muscles through it (Figure 2B). The aneurysm was repaired using a Dacron patch with excision of the excess tissue (Figure 1B). The patient came out of the cardiopulmonary bypass, and was able to maintain good haemodynamics during the first postoperative day. However, she developed hypotension on the third postoperative day with a gradual decrease in urine output. Her condition continued to deteriorate despite maximum inotropic support, and the patient expired on the fifth postoperative day.

Microscopic examination of the recovered tissue from the aneurysm showed variable myocyte hypertrophy and interstitial fibrosis. There was marked interstitial infiltrate comprising lymphocytes, histiocytes, and high proportion of eosinophils without any granuloma or caseous necrosis. All these features were in favour of hypersensitive or eosinophilic myocarditis.

Discussion

Submitral aneurysms are rare and have been described for the first time by Abrahams et al. among the black African population.[1] They are diseases of obscure origin with rare and varied causes. Genetic cause has been suggested because of racial predilection.[5] Submitral aneurysm associated with Takayasu's arteritis[6] and tubercular pericarditis[7] has been reported, suggesting the role of infection and inflammation in the pathogenesis of this disease. On the other hand, reports of non-infectious and non-traumatic aneurysms support the notion that the submitral aneurysms result from a congenital defect in the mitral valve ring. This fact is supported by the findings that submitral aneurysms occur only underneath the posterior leaflet of the mitral valve and below the intermediate portion of the left aortic sinus.[8] This is contrary to the occurrence of sinus of Valsalva aneurysms, which can involve any of the sinuses. Additionally, there are foetal echocardiographic evidences to confirm the congenital origin of many of these aneurysms.[9]

The basic pathology in these lesions has been described as a disjunction between the LV musculature and the left atrium-mitral valve region due to the disturbance of complex embryogenesis, which ties up the left atrium, LV and the mitral valve ensuring electrical isolation.[10] In a recent study, Nayak et al. described a submitral membranous curtain as the potential anatomical basis of these aneurysms.[10] This membrane, which extends along varying lengths of the posterior annulus beyond the posteromedial commissure, forms an area of potential weakness. Infection of this inherently strong area may predispose to aneurysm formation.[11] The extent of the involvement in the aneurysmal process can vary from a small area to the entire region of the posterior mitral annulus.[11] Based on this, DuToit et al. classified submitral aneurysm into 3 types, namely Type I—single localised neck; Type II—multiple necks (separate distinct openings); Type III—involvement of the entire mitral annulus.[11]

Patients with submitral aneurysms may be asymptomatic or present with mitral insufficiency with or without LV dysfunction. They may present with myocardial ischaemia secondary to the compression of left main artery,[2] left circumflex artery,[12] thromboembolism,[13] and arrhythmias.[3,14]

The case reported here is a large submitral aneurysm presented in a state of cardiogenic shock. This could be due to

the inadequate stroke volume secondary to stealing of a large part of it into the aneurysm. In addition, inflammatory nature of the origin of the aneurysm was evident by the elevated inflammatory parameters.

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