Multiple angiomatous mitral valve cysts leading to floppy mitral valve syndrome

Floppy mitral valve syndrome can be caused by various abnormalities, including structural abnormalities that can be identified by echocardiography. A spectrum of floppy mitral valve structure has been demonstrated by echocardiography, with mitral regurgitation occurring more frequently in patients with multiple and more severe anatomic abnormalities. In addition to the presence of prolapse and regurgitation, the assessment of leaflet thickness, leaflet length, annular diameter, and chordal length is fundamental to the definition and stratification of patients with mitral valve prolapse associated with floppy mitral valve.

**CASE REPORT**

A 46 year-old woman was admitted with symptoms of palpitation, dyspnea, and angina pectoris on exertion of eight-year history. These complaints were also present for a year on minor and minimal exercise related to housework. No information on personal or familiar cardiac anamnesis could be derived from the patient. On admission, she was evaluated as NYHA class III-IV. On cardiac examination, the apical impulse was weak and an S₃ was heard. P₂ was loud and there was a grade 3-4/6 apical pansystolic murmur. The electrocardiogram showed normal sinus rhythm with P mitrale. There was left ventricular hypertrophy with strain pattern. The two-dimensional echocardiogram revealed multiple cystic-tumoral formations attached to the anterior mitral leaflet, resulting in pansystolic pseudoparachute-like floppy mitral valve prolapse, and severe mitral regurgitation. The patient underwent prosthetic mitral valve replacement following removal of the mitral valve and multiple cystic-tumoral formations. She had an uneventful postoperative course. Histological diagnosis was diffuse angiomatous cystic development of vasculatory tumor-like structures due to verrucous endocarditis.

**Key words:** Echocardiography; heart valve diseases/ultrasonography; mitral valve insufficiency; mitral valve prolapse/pathology/surgery.

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ventricular enlargement with a cardiothoracic ratio of 0.56 on a chest X-ray.

Echocardiography revealed multiple cysts attached to the anterior mitral leaflet, pansystolic pseudoparachute-like floppy mitral valve prolapse, and severe mitral regurgitation (Fig. 1). The anterior and posterior mitral leaflets were enlarged and deformed. The patient could not be stabilized clinically with digitalis, diuretics, and vasodilators, and she was submitted to open heart surgery. Multiple cystic-tumoral formations were observed with the removal of the mitral valve (Fig 2). Diffuse tumor-like multiple cysts of varying size originated from the mitral valve tissue. The pulmonary artery was slightly dilated. There was global cardiomegaly. The intracardiac tumoral-cystic formation protruded into the left atrium at surgical exploration and gross anatomopathologic examination. One of multiple cystic formations was giant, bloodsucker-shaped, and of saccular formation. The mitral valve was replaced with a Björk-Shiley 33-mm prosthetic valve and the patient had an uneventful postoperative course. Myocardial hypertrophy was determined in the left atrial appendage. The suspected diagnosis was histologically confirmed as diffuse angiomatous cystic development of vasculatory tumor-like structures due to verrucous endocarditis.

**DISCUSSION**

Mitral prolapse is a parachute-like protrusion of the valve into the left atrium. The floppy mitral valve prolapses in such a dynamic manner that it becomes a space-occupying lesion within the left atrium. Prolapsing floppy mitral valve also causes mitral valvular regurgitation.\(^1\)\(^-\)\(^3\) With the onset and gradual progression of mitral valvular regurgitation, alterations in the size and performance of the left atrial and ventricular chambers occur, resulting in left atrial and ventricular myopathy.\(^1\) Typically, prolapsed mitral valve tissue is shiny, grey-white edematous, and contains increased mucopolysaccharides in the zona spongiosa and chordae tendinae, and exhibits myxoid balloon cell degeneration (floppy mitral valve syndrome); however, in our case, these histopathologic features were not observed and the specimens exhibit-
Blood cysts are congenital cysts located in the endocardium, particularly along the lines of closure of heart valves. They are lined by flattened endothelium and filled with nonorganized blood. Mitral valve blood cysts associated with floppy mitral valve are an extremely rare entity. They can be formed by trapping of blood in sacculated dissolved collagenous fibrous layers. If they cause functional derangement of the mitral valve, they must be excised surgically with or without prosthetic valve replacement. In our case, the floppy mitral valve was removed surgically along with the multiple cysts and was replaced with a prosthetic valve.

Mitral valve blood cysts can also be associated with mitral valve hamartoma. Hamartoma is a benign tumor-like nodule composed of an overgrowth of mature cells and tissues that are normally present in the affected tissue, but shows disorganization often with one element predominating. There are angiomatous subtypes, but hamartoma is usually a solid mass such as a cardiac rhabdomyoma. Our case also differed from mitral valve hemangioma or hamartoma histopathologically.

In conclusion, this case is a striking echocardiographic example of multiple cysts associated with floppy mitral valve, which was further confirmed by macroscopic and microscopic diagnosis as a tumor-like cystic lesion secondary to verrucous endocarditis. In addition, a cystic-tumoral lesion of angiomatous subtype arising from verrucous endocarditis associated with floppy mitral valve syndrome is an extremely rare entity causing severe mitral regurgitation and requiring mitral valve surgery.

REFERENCES