Idiopathic mitral valve prolapse with tricuspid, aortic and pulmonary valve involvement: An autopsy case report

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ABSTRACT
Mitral valve prolapse (MVP) is usually asymptomatic, but can be associated with complications such as infective endocarditis, mitral regurgitation, thromboembolism and sudden cardiac death. It has been very rarely reported to occur in association with other valvular involvement. A 55-year-old male patient was brought dead and at autopsy the mitral valve orifice was stenotic and the leaflets were enlarged, myxoid and bulging suggestive of MVP and chordae tendinae were thickened, stretched and elongated. Similar changes were seen in the tricuspid valve. The pulmonary and aortic valves also showed myxomatous degeneration of their cusps. Myxomatous degeneration is the most common cause of MVP and it can be associated with involvement of the other valves. Concomitant involvement of the aortic valve has been reported, however it is very rare and simultaneous involvement of the pulmonary valve has not been reported in the literature so far. We report a case of MVP associated with myxomatous degeneration of the tricuspid, pulmonary and aortic valves.

KEY WORDS: Aortic, mitral valve prolapse, pulmonary, tricuspid

INTRODUCTION
Mitral valve prolapse (MVP) is an abnormal displacement of the mitral valve into the left atrium during systole. It is usually asymptomatic but can be associated with complications like infective endocarditis, mitral regurgitation, thromboembolism and sudden cardiac death.[1] MVP has been very rarely reported to occur in association with tricuspid and aortic valve involvement.[2] However, pulmonary valve involvement has not been reported with MVP. We report here a case of mitral and tricuspid valve prolapse associated with myxomatous degeneration of the aortic and pulmonary valves.

CASE REPORT
A 55-year-old male, chronic alcoholic and hypertensive was brought dead to our hospital. At autopsy, significant findings were seen in the heart and brain. The heart was globular and weighed 300 gms. The mitral valve orifice was stenotic, the leaflets were enlarged, myxoid and bulging towards the left atrium with interchordal hooding suggestive of MVP [Figure 1]. The chordae tendinae were thickened, stretched and elongated and there was a small focus of calcification in the posterior mitral leaflet [Figure 2]. Similar changes were seen in the tricuspid valve [Figure 3]. The pulmonary and aortic valves also showed myxomatous degeneration of their cusps [Figures 4 and 5]. There was left atrial dilatation and mild hypertrophy of the left ventricle.

DISCUSSION
The prevalence of MVP is variable. The worldwide prevalence of MVP is between 0.4% and 35% whereas Indian prevalence by echocardiography studies is between 2.7% and 16%.[3,4] This wide prevalence could be due to the variety of populations studied, both hospital-based and healthy volunteers with a minority still being unrecognized since most are usually asymptomatic. There are very few autopsy studies on MVP with a reported incidence of about 4%-5% at autopsy.[5,6]

MVP is more common in females as compared to males.[3] Myxomatous degeneration is the most common cause of MVP in which there is excessive accumulation of glycosaminoglycan.
Desai and Amonkar: Idiopathic mitral valve prolapse

material within leaflets and cusps. It can be associated with diseases of tricuspid (commoner) and aortic (less common) valves. Other less common causes include dysfunctional papillary muscles, ruptured chordae tendinae or papillary muscles.[7]

The weight of the heart is increased only in symptomatic MVP but usually remains normal in asymptomatic patients.[8] According to Edwards,[7] the pathological criteria to label the mitral valve as MVP include interchordal hooding (the free margins of the mitral valve leaflet bulges between the chordal insertions) which should be more than 4 mm in height and should involve half of the anterior leaflets or more than 2/3rd of the posterior leaflet of the mitral valve. Whenever the leaflets are voluminous with marked bulging into the left atrium, MVP is usually of a long duration which was seen in our case.

On gross examination of the valve, the deposits can be focal/ generalised. With generalised myxomatous degeneration, both atrioventricular and semilunar valves can be affected as was seen in our case. Semilunar valve cusps are thinner and translucent which makes them prone to aneurysm and rupture.[9] The spongiosa layer of the rough zone is the main site of myxomatous change in MVP.[5]

Patients of MVP with generalized myxomatous degeneration can have simultaneous involvement of other valves as seen in our case where MVP was seen to be associated with tricuspid, aortic and pulmonary valve prolapse. Kasper et al.[9] studied 27 patients with tricuspid valve prolapse and found 8 patients (29.6%) to have concomitant mitral and tricuspid valve prolapse which was statistically significant ($P < 0.02$). Concomitant involvement of the aortic valve has been reported, however it is very rare and simultaneous involvement of the pulmonary valve has not been reported in the literature so far.[2]

MVP can be associated with complications like infective endocarditis, mitral regurgitation, thromboembolism and sudden cardiac death. Mitral regurgitation is the most common complication of MVP. Sudden death in MVP occurs in less than
Desai and Amonkar: Idiopathic mitral valve prolapse

1% of cases; usually occurring secondary to ruptured chordae tendinae and mitral valve leaflets. Arrhythmias can occur but are rare.

In our case, we had a stenotic mitral valve, which is extremely rare in MVP. Furthermore, to the best of our knowledge, this is the first reported case of quadruple valve involvement in association with MVP.

REFERENCES


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