Isolated Myxomatous Flail Aortic Valve Causing Severe Aortic Regurgitation

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A 60 years old lady presented to our institute with complains of progressive dyspnea on exertion, increasing from New York Heart Association functional class II to class III over last 2-3 months. She did not have any history of fever, chest pain, palpitations, pre-syncpe or syncope.

On examination, her pulse rate was 90 per minute regular and the blood pressure was 130/60 mm Hg. Rest of the general physical examination was unremarkable with no clinical features suggestive of Marfan’s syndrome, rheumatic heart disease or infective endocarditis. Cardiovascular examination revealed a long diastolic murmur in left upper and mid parasternal region.

Her transthoracic 2D-echo revealed tri-leaflet aortic valve with flail right coronary cusp which was prolapsing into left ventricular outflow tract (Figure 1) causing severe aortic regurgitation. The aortic annulus (22 mm) and the root (38 mm) were of almost normal sizes (Figure 2) and there was no evidence of aortic dissection. Other valves were structurally normal. The left ventricular size was increased but the systolic function was preserved. There was no evidence of infective endocarditis.

Among her first-degree relatives, none had any major cardiac illness. The clinical examination was also unremarkable with none of them having marfanoid features.

In view of the clinical symptoms and echocardiographic features, the patient underwent aortic valve replacement. Histopathological examination of the resected aortic leaflet tissue revealed myxomatous degeneration (Figure 3).

Flail aortic leaflet secondary to myxomatous degeneration is a relatively less common cause of aortic regurgitation but almost invariably occurs in association with some other abnormality such as bicuspid aortic valve, aortic root dilatation, aortic dissection, aortic valve endocarditis, subaortic ventricular septal defect or only the marfanoid habitus.¹⁻³ In our patient, however, none of these were present and the flail aortic leaflet was the only abnormality found. There was no evidence of Marfan’s syndrome or Marfanoid habitus in the family also. This type of presentation is extremely rare with very few cases reported in the literature so far.³⁻⁵

References

regurgitation: a clinico-morphologic study. *Int J Cardiol* 1994; 45:129-134
