Congenital Aortic Regurgitation due to Aortic Valvar Fenestration with Associated Aortic Dissection

Sir,

Congenital aortic regurgitation (AR), in general, is rare and that produced by the fenestrations of semi-lunar cusps of the aortic valve is hardly ever reported. Fenestrations are deficiencies of valvar tissue, located close to the cuspal free margins. As they do not extend beyond the lines of closure, they are of little clinical importance. We came across a young adult in which these fenestrations produced AR with associated aortic dissection.

A 30-year-old male, chronic smoker and alcoholic, was admitted for wrenching chest pain, radiating to the back and the lower abdomen and tingling numbness in the right lower limb. Prior to these symptoms, he used to have episodes of breathlessness and palpitations, for which he never sought medical therapy. On examination, the general condition was poor. On the right side, the brachial and femoral arteries were weakly pulsatile while the other arteries were not felt. The findings of mild superior mediastinal widening on chest radiography, and mild pericardial effusion, dilated aortic root and ascending aorta with 45% left ventricular ejection fraction on two-dimensional echocardiography, substantiated the clinical diagnosis of acute aortic dissection. There were no Marfanoid features. The patient expired before surgical intervention.

At autopsy, there was marked enlargement of the heart (540 g) with moderate left ventricular dilatation and hypertrophy. There was mild aortic annular dilatation. Large fenestrations (0.5-0.8 cm) were seen at the commissural aspects of the right and non-coronary cusps, which extended towards their bases (Figure 1). Similar smaller deficiencies were also seen adjoining the commissure between coronary cusps. The rounding of the free margins of the cusps with prominent central nodules indicated the presence of AR. The ascending aorta was aneurysmally dilated with two intimal tears, one 2 cm transverse at the sino-tubular junction above the right and non-coronary cusps; and the other circumferential, situated 1.5 cm above the sino-tubular junction (Figure 1). This resulted in type 1 aortic dissection, extending into the proximal portions of the arch arteries, right subclavian artery and common iliac arteries. Other arteries were spared. The dissection had ruptured into the pericardial space. Histologically, the entire aorta revealed many areas of cystic medionecrosis.

Fenestrations are caused by age-related degeneration and are hence, common in the elderly. At most times, AR results with the rupture of the bridging strand or incorporation of the fenestrations at the line of coaptation seen with aortic annular dilation or solely due to large-sized fenestrations. However, none of the above cases reported or reviewed were young, and also did not have associated aortic dissection. In the reported case, there were large fenestrations with obvious anatomic features of regurgitation and prominent foci of cystic medionecrosis in the entire aorta. Since valve development and acquisition of elastic fibres in the aortic wall occurs almost at the same time, we assume a congenital developmental error in our case that produced concomitant degenerative changes in the cusps with large fenestrations, and medial degeneration with acute dissection.

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References


Figure 1: Large fenestrations (arrows), affecting all cusps of aortic valve (av) with intimal tears (T1, T2) and aortic dissection. Note rolling of the free margins and shortening of the cusps.