Rupture of Aortic Valve Secondary to Aneurysm of Ascending Aorta

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Rupture or perforation of an aortic cusp usually may be related either to bacterial endocarditis or to trauma. In that type of aortic valvular insufficiency which is associated with dilatation of the ascending aorta, the aortic valve is, as a rule, intrinsically normal, while mechanical distortion of it by the dilated aorta causes it to function improperly.

The case that forms the subject of this report illustrates an unusual condition wherein an aneurysmal ascending aorta caused distortion of two aortic cusps with eventual rupture of one and gross fenestration of the other.

The patient in whom these observations were made was a 49-year-old man whose fundamental clinical problem was that of severe aortic insufficiency (fig. 1). The patient's body build, eyes, and hard palate were not remarkable. The history and serologic tests of serum and of spinal fluid for syphilis gave negative results. There was neither a history of trauma nor a past illness that might have represented bacterial endocarditis. Death resulted from cardiac failure before any surgical therapy was attempted.

At necropsy, the following observations were made. Beginning at the aortic sinuses and involving the major part of the ascending aorta, there was gross aneurysmal dilatation. A sharp line separated the diseased part of the aorta from the grossly normal vessel beyond (fig. 2). The fundamental process leading to the aortic lesion appeared to have been cystic medial necrosis of the aorta with secondary "incomplete rupture" of the aortic wall (fig. 3). As the edges at the rupture recoiled one from the other, the greater part of the ascending aorta was left devoid of media. This weakened segment not only dilated but elongated in a supra-inferior direction, causing the attached aortic cusps to be distorted. The process of distortion of the cusps had two consequences. One involved stretching of the cusps in a side-to-side direction, with bowing of them. The other caused the cusps to be lengthened upward as the ascending aorta became elongated.

Tension upon the cusps in the directions cited caused the posterior cusp to become markedly attenuated and finally to develop a wide (about 2 cm.) fenestration. In the posterior aspect of the left cusp, the same process caused rupture (fig. 4, Left). No evidence for syphilitic or other forms of aortitis was encountered.

Of additional interest is the fact that the aneurysmal ascending aorta showed extensive atherosclerosis, a process that commonly occurs secondarily in aneurysms regardless of etiology. Removed from the aneurysm only minimal aortic atherosclerosis was present.

The calcification in the atherosclerotic lesions in the aneurysm, although extensive and readily demonstrable in the specimen (fig. 4, Right), had not been identified in the plain roentgenograms made during the life of the patient. Nevertheless, it is recognized that, with somewhat different exposure, this process might have been observed. This leads to the observation that, although syphilitic aortitis remains the most common cause of extensive...
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Upper. Selective aortogram made with tip of the catheter in the ascending aorta. Upper Left. Anteroposterior view. There is marked dilatation of the ascending aorta (A) and evidence of regurgitant filling of the left ventricular cavity (LV). Upper Right. Lateral view. The prominent dilatation of the ascending aorta and evidence of massive aortic insufficiency are clearly evident. Plain roentgenography had shown evidence of marked cardiac enlargement and congestion of the lungs. The electrocardiogram showed signs of left ventricular hypertrophy. Lower. Phonocardiogram made over the right second intercostal space in a parasternal position. This shows a prominent diastolic murmur of diminuendo type.

Figure 1

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Figure 2
Left. The left ventricle and thoracic and upper abdominal portions of the aorta. In addition to left ventricular enlargement, there is a lesion involving the ascending aorta. This is characterized by pronounced dilatation of this segment of the vessel. Bordering this area with the intact aorta is a sharp line (arrows) representing the retracted upper edge of the "incomplete rupture" of the aorta. Right. Close-up view of the aortic valve and adjacent structures. The irregular lining of the aneurysmally dilated ascending aorta results from atherosclerotic lesions that developed secondarily to the presence of the aneurysm. The sharp upper edge of the aneurysm represents the upper extremity of an "incomplete rupture" of the aorta. After retraction of the edges of the rupture (arrows), the major part of the ascending aorta was left devoid of significant medial tissue. The left extremity of the posterior aortic cusp (P.) has been stretched upward by the process of elongation of the ascending aorta. The posterior extremity of the left aortic cusp (L.) has also been elongated upward and it has ruptured (close-up in fig. 4, Left).

Figure 3
Photomicrographs of aorta. Left. The thoracic aortic wall removed from the area of aneurysmal dilatation showing small areas of cystic medial necrosis and one zone of loss in continuity of the media with retraction of elastic tissue from this zone. The picture is compatible with that seen in idiopathic dilatation of the ascending aorta and in the Marfan syndrome. Although the patient, who is the subject of this report, did not manifest outward signs of the Marfan syndrome, he may represent an example of forme fruste of that condition. It is considered that the fundamental weakness of the media portrayed in this illustration was responsible for a gross break in continuity of the intima and media, which in turn led to the aneurysmal change of the ascending
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Figure 4
Left. Close-up view of the adjacent parts of the posterior (P.) and the left (L.) aortic cusps. The left upper extremity of the posterior aortic cusp (arrow P.) has been carried upward by the elongation of the ascending aorta and a large fenestration has appeared in this cusp. The upper extremity of the ruptured left aortic cusp is represented by a nipple-like protrusion from the wall of the ascending aorta (arrow L. U.). The lower part of the ruptured left aortic cusp appears as a strand pointing upward (arrow L. L.). The left cusp is now prolapsed. The wall of aneurysmally dilated ascending aorta is in the background and is grossly irregular by virtue of secondary atherosclerotic deposits. Right. Roentgenogram of specimen of the heart and ascending aorta. There are numerous deposits of calcium (arrows) in the wall of the aneurysmally dilated ascending aorta.

Figure 3 (Continued)
aorta. Elastic tissue stain; × 57. Right. Photomicrograph of the junction of the intact aortic wall and the aneurysmally dilated ascending aorta. Abrupt loss in continuity of the aortic media (arrows) at the junction of the intact aorta (right side of illustration) with the aneurysm (left side of illustration). The sharp edge between the aneurysmal and nonaneurysmal parts of the aorta shown in figure 2 is represented by the break in continuity of the media, which is shown in this photomicrograph. Elastic tissue stain; × 50.

Summary
A case of aneurysm of the ascending aorta, the result of cystic medial necrosis, is presented. The feature of peculiar interest is that as the diseased part of the aorta became elongated it stretched the attached aortic cusps. As a consequence, one cusp developed a gross fenestration, while the other ruptured and prolapsed.
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