CLINICOPATHOLOGIC CORRELATIONS

Aneurysms of the Thoracic Aorta Complicating Coarctation

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SUMMARY
Coarctation of the aorta may become complicated by aortic aneurysms involving either the proximal or distal compartment of the vessel. Such aneurysms principally are consequences either of hypertension or of bacterial infection. The aneurysms in either compartment may be of the classical dissecting or saccular types. Saccular aneurysm may result from infection or laceration of the aorta without intramedial dissection of blood.

In the ascending aorta, an additional basis for aneurysm is aortic stenosis, the latter either being congenital, or more commonly, acquired calcific in a congenital bicuspid valve.

Additional Indexing Words:
Dissecting aneurysm  Incomplete dissecting aneurysm  Mycotic aneurysm  Aortic stenosis

Among patients with coarctation of the aorta, a complication may develop or be manifest in the form of an aortic aneurysm. Although this possibility exists in children, such lesions are usually not observed until adolescence or adult life. Included among such aneurysms is the false aneurysm which results from leakage at the anastomosis done as part of the surgical correction for the fundamental lesion.1,2 This particular condition will not be further considered, as emphasis will be made upon those aneurysm which occur spontaneously. Usually, such aneurysms are derived either from 1) hypertension, which characteristically is present, along with varying degrees of cystic medial necrosis, or 2) from the complication of infection. An additional potential is that aneurysm of the ascending aorta which may complicate the aortic stenosis which forms on a bicuspid valve.

Infection may occur at one or two sites, namely, the commonly present congenital bicuspid valve or the lining of the descending aorta at the site of...
impact of the jet-like stream which passes through the narrow segment of the vessel. Aneurysms may involve either the compartment proximal or that distal to the coarctation.

The most common basis for an aneurysm is hypertension and complicating spontaneous laceration of the aortic intima and varying depth of the underlying media. Following the laceration, progressive dissecting of blood within the media of the aorta may occur, the condition commonly called classical dissecting aneurysm of the aorta. If the laceration is followed by limited or no appreciable intramural dissection of blood, the tendency is for a saccular aneurysm to develop at the weakened zone corresponding with the laceration. The resulting aneurysm may be termed an incomplete dissecting aneurysm.

Aneurysms of Proximal Compartment

The most common type of aortic aneurysm complicating coarctation is classical dissecting aneurysm of the proximal compartment\(^6,4\) (fig. 1). The internal tear usually occurs in the ascending aorta and this is followed by external rupture of the ascending aorta leading to hemopericardium (fig. 2). As with all dissecting aneurysms, obstruction of aortic branches may develop by the extension of the aortic medial hematoma into the walls of branches. A characteristic of dissecting aneurysm complicating coarctation, however, is that the intramural dissection classically does not extend beyond that segment of the aorta which harbors the lesion of coarctation, so that only the coronary arteries and branches of the arch are subject to obstruction.

Figure 3

Diagrams of incomplete dissecting aneurysm of the aortic arch complicating coarctation.

Figure 2

Dissecting aneurysm of the proximal aorta complicating coarctation in a young man. a. Roentgenogram of thorax. b. Interior of the aorta shows major laceration which had resulted in dissecting aneurysm and hemopericardium. At the junction of the arch and the descending portions is a characteristic indentation (point of arrow) in the exterior of the aorta corresponding with the zone in which the lumen showed major narrowing.
In subjects with coarctation, those lacerations of the aorta occurring proximal to the coarctation and not followed by intramural extension of blood most often lie in the arch and lead to saccular aneurysm (incomplete dissecting type)⁵ (fig. 3).

Those saccular aneurysms of the proximal compartment which result from infection are uncommon. Such aneurysms are classified as mycotic and result from infection secondary to bacterial endocarditis of an associated congenital bicuspid aortic valve (fig. 4a and b). Mycotic aneurysms usually involve the aorta only a short distance above the valve and they may rupture into adjacent structures such as the right atrium or the outflow tract of the right ventricle leading to an acquired left-to-right shunt.

As bacterial endocarditis of the aortic valve in coarctation patients is classically dependent upon the presence of a congenital bicuspid aortic valve, the potential for an aortic mycotic aneurysm remains in those patients with an anomalous valve, even after repair of the coarctation.

It is recognized that calcific aortic stenosis commonly forms on a congenital bicuspid aortic valve. This means that in a significant number of patients in whom the coarctation has been corrected there remains the chance of aortic stenosis developing with the passage of time. In such patients or in those in whom congenital aortic stenosis coexists with the coarctation, the potential for an aneurysm of the ascending aorta remains, since aortic stenosis may effect a spontaneous laceration of the ascending aorta.⁶ ⁷ Such a

Figure 4

Diagrams of mycotic aneurysm of ascending aorta complicating bacterial endocarditis of a congenital bicuspid aortic valve. a. Vegetations of bacterial endocarditis (S.B.E.) on aortic valve are sources of infection (Veg.) of aortic wall. b. Destruction of aortic tissue leads to a mycotic aneurysm (M.A.) c. Calcific aortic stenosis developing in congenital bicuspid aortic valve with secondary laceration of ascending aorta. The latter process may lead to intramural dissection of blood (D.A.) of varying degree. The process shown may occur years after adequate resection of the coarctation.
Aneurysms related to the aorta distal to the site of coarctation include aneurysmal formation in intercostal arteries, which vessels play a well known role in the collateral circulation. Aneurysms of the descending aorta itself may occur. In that segment, aneurysms, with the exception of that complicating aortic stenosis, are derived from the same causes as those involving the aorta proximal to the obstruction.

Since classical dissecting aneurysm beginning in the descending aorta is distinctly less common than the relatively common dissecting aneurysm of the proximal compartment, isolated cases have been described (fig. 5a). The basis for this uncommon complication is probably hypertension. It is to be recalled that, while in the aorta distal to the coarctation the systolic pressure is within normal limits, diastolic hypertension, occurs here as it does in the proximal compartment.

Saccular aneurysms involving the descending aorta may be part of classical dissecting aneurysm

**Aneurysms of Distal Compartment**

Diagrams of dissecting aneurysm of descending aorta. a. Classical situation. b. The classical dissecting aneurysm has become complicated by a localized saccular aneurysm.

Diagram of incomplete dissecting aneurysm of descending aorta resulting in a saccular aneurysm distal to the site of coarctation.
(fig. 5b) or result either from laceration with limited dissection (incomplete dissecting aneurysm) or infection of the aortic wall.9 Laceration without significant intramedial dissection of blood represents the same primary condition as in classical dissecting aneurysm (fig. 6).

Examination of the mouth of the aneurysm shows an abrupt interruption in the continuity of the aortic media (fig. 7), giving evidence that laceration of the wall had been the basis for the aneurysm.

The other cause for saccular aneurysm involving the descending aorta is infection of the aorta.10,11 The narrow, high velocity stream passing through the zone of coarctation may impact against the arterial wall. A jet lesion may develop at the site of impact and upon such a lesion an infective aortitis may develop. With progress of infection, there is progressive destruction of the aortic wall leading to a mycotic aneurysm (figs. 8 and 9). Infection of the aorta may be the only intravascular site of such involvement or it may be a complication of aortic valvular bacterial endocarditis.

Figure 7

a. Thoracic roentgenogram of a patient with coarctation of the aorta and a saccular aneurysm involving the upper portion of the descending aorta. The aneurysm has displaced the esophagus. b. Photomicrograph of one edge of the mouth of the aneurysm. There is abrupt interruption in the continuity of the aortic media (arrow) representing the laceration which allowed the formation of the saccular aneurysm (A.). Elastic tissue stain (x4.5).

Figure 8

Basis for mycotic aneurysm of descending aorta developing upon the site of impact of the jet-like stream extending through the narrow segment of the aorta. a. The first step appears to be that of the formation of the jet lesion (J.L.). b. This is followed by infection with formation of vegetations (Veg.). c. Finally a mycotic aneurysm (M.A.) results from loss of integrity of the aortic wall on the basis of infection.
Aortic aneurysm distal to coarctation. The lesion is presumed to have resulted from infection of the aorta. a. Roentgenogram of thorax. b. The opened aorta from in front. Beyond the zone of coarctation (Coar.) is a circular perforation (between arrows) in the medial aspect of the upper descending aorta. This led to an aneurysm which ruptured into the right pleural cavity. c. The tracheal bifurcation and the upper descending aorta viewed from behind. The fundus of the aneurysm has been removed. The probe extends from the aortic lumen through the ostium observed in b and enters the aneurysm (A.M.).
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