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Nonneoplastic Diseases of Aorta and Pulmonary Trunk—Part IV

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This five-part review focuses on selected nonneoplastic diseases of the aorta and pulmonary trunk. Because many more diseases affect the aorta compared with the pulmonary trunk and right and left main pulmonary arteries, most of this review will be devoted to disorders of the aorta. Part IV of the series discusses cystic medial degeneration, trauma, and congenital diseases of the aorta.

Key words: aorta, dissection, Marfan's syndrome, cystic medial necrosis, sinus of Valsalva

Cystic Medial Degeneration

Cystic medial necrosis is a term first used by Erdheim in 1929[1,2] as a modification of the term medionecrosis described by Gsell.[3] These terms describe a process of medial elastic fiber degeneration and deposition of acellular basophilic material. In mild forms of this condition, the elastic elements of the media are not noticeably affected.[4] With more basophilic deposition, medial fibers are interrupted and retract from sites of interruption. The areas of medial interruption produce acellular "cyst-like" spaces. In advanced forms of the disease, extensive areas of the elastic layers are interrupted (Fig. 1). These patients probably have a congenital abnormal media and have a change similar to that observed in patients with Marfan's syndrome. Lesser degrees of this condition are probably acquired and may occur in response to hemodynamic stresses as present in systemic hypertension,[5] aortic valve stenosis,[6] and bicuspid aortic valve.[7] Schlatman and Becker[8] recently showed that certain amounts of elastic degeneration occur with normal advancing age. The extensive cystic medial degeneration with interrupted medial layers account for the diffuse dilation of the aorta as well as intimal tears that occur in patients with Marfan's disease.

Trauma

External blunt trauma to the thorax may cause laceration of the aorta.[9–22] Two anatomic sites are particularly susceptible: the junction of the arch and the descending aorta, and the proximal part of the tubular segment of the ascending aorta.[5] The latter site is more common and has the potential for hemothorax or aneurysm formation. Traumatic rupture of the ascending aorta may cause either hemopericardium or prolapse of aortic valve cusps with secondary pure aortic regurgitation.[5]

Congenital Diseases of Aorta

Certain nonneoplastic disease of the aorta are congenital in origin: supravalvular aortic stenosis, coarctation of the aorta, aortic arch interruption, and sinus of Valsalva aneurysm.

Supravalvular Aortic Stenosis

Supravalvular aortic stenosis is characterized by an obstructive anomaly in the tubular portion of the ascending aorta. Three anatomic variants are recognized: hourglass type, hypoplastic type, and membranous type.[23] Occasionally, surgical specimens are submitted from patients with supravalvular aortic stenosis, including excised aortic membranes and/or excision of the narrowed segment. This condition may be associated with stenosis of branches of the aortic arch and pulmonary arteries.[24,25]

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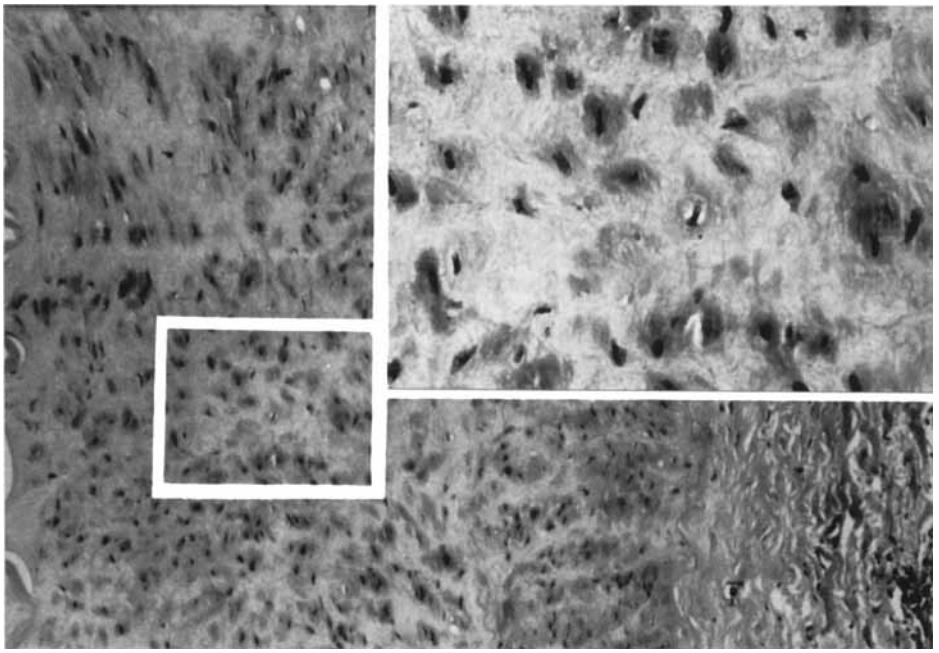

FIG. 1 Idiopathic medial degeneration in a patient without Marfan's syndrome. From Ref. No. 1 with permission.

Coarctation of the Aorta

Coarctation of the aorta (Fig. 2) is characterized by a localized deformity of the aortic media.[5] The process takes the form of a "curtain-like thickening" that protrudes into the lumen from the anterior, posterior, and superior aspects of the aortic wall.[26] The curtain-like thickening encroaches on the aortic lumen and causes it to be eccentric and narrow. The usual position is beyond the origin of the left subclavian artery and near the aortic entrance of the ductus arteriosus. Coarctation of aorta is coupled with dilation of the vessel proximal to the lesion and distal to the lesion, yielding in some instances a figure-3 pattern.

Glancy *et al.*[27] studied 70 excised, serially sectioned coarctation segments from 70 patients: occluded in 4, up to 0.5 mm internal diameter in 22, from 0.6 to 2 mm in 26, from 2.1 to 5 mm in 14, and >5 mm in 4 patients. The most significant anatomic factor causing the coarctation was invagination of the media from the posterior aortic wall, but intimal proliferation ("jet lesion") at and immediately distal to the invagination contributed to the narrowing.

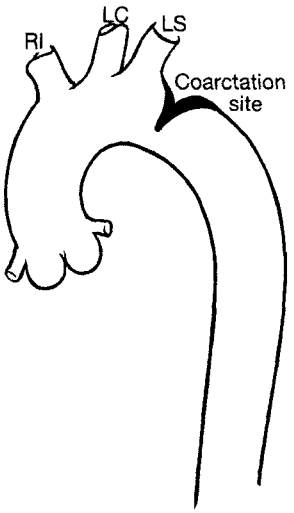

FIG. 2 Diagram shows coarctation of aorta. In many young patients, the coarctation is associated with a patent ductus arteriosus. RI = right innominate, LC = left cartoid, LS = left subclavian. From Ref. No. 1 with permission.

Sinus of Valsalva Aneurysms

Edwards and Burchell[28, 29] postulated that a congenital deficiency in the elastic continuity between the aorta and aortic valve ring predisposed the area to the development of a sinus of Valsalva aneurysm. The most common location of the aneurysm is the right sinus of Valsalva, followed by the posterior, then left sinus. The aneurysms enlarge in response to aortic pressure, and the direction of expansion is determined by the area of least resistance; thus, most aneurysms rupture into the right side of the heart.

Aortic Arch Interruption

In interruption of the aortic arch, it is the luminal continuity between the ascending and descending aorta that is interrupted. In some instances, the aortic segments are completely in-

terrupted; in others, a fibrous stand (no lumen) connects the two portions of the aorta. Depending on the site of luminal interruption, there are three basic types: A, B, and C.[30] In type A, the interruption is between the left subclavian artery and the ductus; in type B, it is between the left subclavian and the left common carotid arteries; and in type C, it is between the left common carotid and the innominate artery. The letters A, B, and C also reflect the respective frequency of the types of arch interruptions (in decreasing order).

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