Clinical Pathologic Correlations

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Nonneoplastic Diseases of Aorta and Pulmonary Trunk—Part III

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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 III of this five-part series discusses the etiology of aortic aneurysms and aortitis.

Key words: aorta, aortic aneurysm, aortitis

Etiology of Aortic Aneurysms

The etiology of aortic aneurysms can be related to their locations.¹ In the abdominal aorta, the classic fusiform aneurysm is of atherosclerotic origin and is by far the most common type of aneurysm in all locations. Unless there is disease of the underlying media, such as syphilis, the ascending aorta is not a site of atherosclerotic aneurysm formation. When the arch and descending aorta are sites of aneurysm of atherosclerotic origin, there usually is aneurysmal involvement of the abdominal portion of the aorta as well. Isolated atherosclerotic aneurysms of the thoracic aorta are rare.²

In the sinus portion of the aorta, aneurysms are either congenital (usually one sinus) or secondary to infection (mycotic, usually one sinus). Diffuse dilation (fusiform) of the ascending aorta occurs in cystic medial degeneration (involves all three sinuses), such as in Marfan's syndrome. Aneurysms of the tubular portion of the aorta may result from chronic aortitis (such as syphilis) (Figs. 1–3) or from laceration (trauma) of the aorta. In spontaneous laceration, aortic dissection may re-

sult.³ Of the various segments of the thoracic aorta (ascending, arch, descending), the arch is the least common site for saccular aneurysm. The two principal etiologies of saccular aneurysms of the arch are chronic aortitis and laceration (spontaneous traumatic). The most common site for a saccular aneurysm of the thoracic aorta is at the junction of the arch and the descending portions. Chronic aortitis or a primary mycotic aneurysm may be the basis for the aneurysm in this location; however, laceration (either spontaneous or traumatic) is the most common etiology at this site.^{1, 2} Aneurysms involving both the thoracic and abdominal portions of the aorta in continuity (thoracoabdominal) are usually atherosclerotic in origin. Periaortic cysts may rarely mimic an aneurysm on an x-ray. Large lymphocysts (Fig. 4) were observed at necropsy in a patient who previously had undergone resection of an abdominal atherosclerotic aneurysm. The lymphocysts resulted from thoracic duct obstruction.

Aortitis

Recognition of aortitis as an entity dates back to the sixteenth century with the recognition of syphilis. The grossly wrinkled intima of the aorta described as having a "tree-bark" appearance was accepted as pathognomonic of syphilis (Figs. 1–3). During the early part of the twentieth century, the association of aortitis with other systemic diseases (rheumatic fever,³ arthritis⁴) began to gain recognition. The "tree bark-ing" gross appearance of the aortic intima was seen with various causes of aortitis in addition to syphilis. In 1937, giant cell arteritis was described in the aorta,⁵ and subsequently other systemic diseases associated with aortitis were recognized: ankylosing spondylitis,⁶ scleroderma,⁷ relapsing polychondritis,⁸ Bechet's syndrome,⁹ Reiter's syndrome,¹⁰ and systemic lupus erythematosus.¹¹

In most forms of aortitis, the inflammation is limited to the media and adventitia without involvement of the intima (except in acute rheumatic fever aortitis¹¹). In the healing phase or chronic phase of aortitis, the cellular infiltrate resolves and the damaged tissue is replaced with collagen. Subsequent retraction of the collagen results in wrinkling and a "tree-bark" appearance. This appearance of the intimal surface of the aorta is characteristic of most forms of aortitis irrespective of the etiology.¹¹

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FIG. 1 Syphilitic aneurysm. (A) Multiple aneurysm (*) of ascending, arch, and descending thoracic aorta from a patient with known syphilis. Atherosclerotic plaque lines the aneurysms. (B) Ascending aorta showing intimal tree-barking indicative of aortitis. The aortic valve is also thickened. From Ref. No. 1 with permission.

Syphilitic Aortitis

Characteristically, syphilitic aortitis involves the proximal aorta and does not extend beyond the renal arteries. The aortic wall is thickened, with maximal thickening of the adventitia and intima. Histologically, a perivascular infiltrate consists of lymphocytes and plasma cells around the vasa vasorum of the adventitia and media (Figs. 1–3, 5). The inflammation

causes destruction of the elastic fibers and smooth muscle cells. In addition, there is severe endarteritis obliterans. Rarely, spirochetes may be recognized in the microorganisms.¹²

FIG. 2 Syphilitic ascending aortic aneurysm. From Ref. No. 1 with permission.

FIG. 3 Syphilitic aortitis. Histologic composite showing focal medial scars and mononuclear cells surrounding vasa vasorum. From Ref. No. 1 with permission.

FIG. 4 (A) Repair of abdominal aortic aneurysm (AAA) with intra-aneurysmal graft (G). (B and C) On the posterior wall, large lymphocysts were present, indicating proximal lymphatic obstruction. From Ref. No. 1 with permission.

FIG. 5 Histologic sections of ascending aorta showing perivascular lymphocytic and plasma cell infiltration in adventitia (A and B), and thickened vasa vasorum (A and B), and perivascular cuffing by chronic inflammation (C) with endarteritis obliterans (D-F). From Ref. No. 24 with permission.

FIG. 6 View of aortic valve from a patient with pure aortic regurgitation from syphilis. The aortic valve (AV) cusps are focally thickened, and the sinus portion of the wall of aorta is thickened. LAD = left anterior descending, LC = left circumflex, R = right coronary arteries. From Ref. No. 1 with permission.

In addition to aortitis, syphilis can cause aortic aneurysms, aortic valvulitis or aortic root dilation with pure aortic regurgitation (Figs. 6, 7), and coronary ostial stenosis (Fig. 1).

Rheumatoid Arthritis

Rheumatoid nodules of rheumatoid arthritis may occur in the aorta,¹³ or a nonspecific panaortitis mimicking syphilis is apparent.¹⁴ Of 38 patients with rheumatoid arthritis studied by Robinowitz *et al.*,¹⁵ 3 had rheumatoid nodules identified in the walls of the aorta. In each patient, the lesions occurred in the proximal portion of ascending aorta.

Ankylosing Spondylitis

Ankylosing spondylitis is an idiopathic inflammatory disorder characterized by progressive bilateral sacroiliitis. In this disease, the aorta is affected in a manner similar to syphilis but its aortic distribution is strikingly different from syphilis.^{1, 16-18} In contrast to syphilis, ankylosing spondylitis involves the aortic valve cusps, causing them to thicken. In ankylosing spondylitis, the adventitial process in the aorta extends below the aortic valve into the ventricular septum (may involve the cardiac conduction system) and down to the anterior leaflet of mitral valve. The subaortic fibrous thickening has been termed a bump.^{17, 18}

Reiter's Syndrome

Reiter's syndrome is characterized by a triad of urethritis, conjunctivitis, and polyarthritis, especially in young men. Aortitis with aortic root dilation may be found in 2 to 5% of patients. The cardiovascular involvement in Reiter's syndrome is similar to ankylosing spondylitis, with prominent involve-

FIG. 7 Diagram compares the various diseases affecting the ascending aorta. From Ref. No. 1 with permission.

ment of the ascending aorta, disruption of elastic tissue, and an inflammatory infiltrate.¹⁹

Relapsing Polychondritis

This condition is characterized by intermittent inflammation of cartilaginous structures of the ear, nose, joints, larynx, and trachea. Pearson *et al.*²⁰ recognized aortic disease as a complication of this disorder. Aortic aneurysms with and without rupture have been reported in 10% of cases.²⁰ The aorta has focal loss of glycosaminoglycans, destruction of the media with mononuclear infiltrate, and a decrease of elastic tissue and sulfated acid mucopolysaccharides. These lesions are distinct from Erdheim's cystic medial degeneration; the loss of glycosaminoglycans and the inflammatory reaction have been considered the result of the release of lysosomal enzymes from damaged cells.²⁰

Giant Cell Arteritis

Giant cell arteritis is characterized by focal granulomatous inflammation of small to medium sized arteries, especially temporal arteries (Fig. 8). The aorta may be involved and grossly appears as wrinkled tree-bark. The lesions are mainly confined to the media (including the intimal-medial junction) with fragmentation of the elastic layers, smooth muscle cell loss, and medial scarring. Giant cells, lymphocytes, and plasma cells are seen in the acute phase. Giant cell aortitis in the healed phase is associated with aneurysm formation. In the healing stages, the aorta has morphologic findings similar to those of ankylosing spondylitis, Takayasu's disease, rheumatoid arthritis, and Reiter's syndrome. For this reason, Marquis *et al.*²¹ have combined these conditions under the term idiopathic medial aortopathy and arteriopathy. Giant cell arteritis can be distinguished from Takayasu's disease,

FIG. 8 Histologic composite of aorta from a patient with giant cell aortitis. From Ref. No. 1 with permission.

although histologically and clinically there is overlap. Takayasu's disease occurs in young females and mainly involves large vessels, whereas giant cell arteritis occurs almost exclusively in persons >50 years of age and involves mainly extracranial vessels.

Bacterial and Fungal Aortitis (Fig. 9)

Infection of the aortic wall by bacteria or fungi usually involves the ascending aorta as a complication of aortic valve infective endocarditis.^{1, 2} Rarely, the process can involve any

FIG. 9 Abdominal aortic aneurysm with fungal infection (zygomycosis) in thrombus. From Ref. No. 1 with permission.

portion of the aorta in the absence of endocarditis as a complication of septicemia (*Salmonella*^{22, 23}). Suppurative aortitis in the absence of endocarditis is called primary suppurative aortitis, and the resulting mycotic aneurysm is called primary mycotic aneurysm.² The aortic wall becomes damaged, with abscess and aneurysm formation. Spread of the infection from the aorta to adjacent tissues (organs, vessels) may lead to shunt formation.

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