Clinical Pathologic Correlations

Nonneoplastic Diseases of Aorta and Pulmonary Trunk—Part I

BRUCE F. WALLER, M.D.,*† JAMES D. CLARY, P.A.-C,† TODD ROHR, P.A.-C†

*Cardiovascular Pathology Registry, St. Vincent Hospital; †Nasser, Smith, Pinkerton Cardiology Inc., Indianapolis, Indiana, USA

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 I of this five-part review discusses general concepts of aorta anatomy and aortic dissection.

Key words: aorta, aneurysm, dissection

Introduction

The aorta is subject to several acquired and congenital diseases as well as to normal aging changes. Acquired nonneoplastic diseases of the aorta include atherosclerosis, dissection, aneurysm, acute and chronic aortitis, medial degeneration with and without Marfan's syndrome), and trauma. These conditions alter the aorta by thickening or thinning, widening or narrowing, tearing or rupturing, or various combinations.^{1,2} Of the acquired conditions, atherosclerosis, dissection, and aneurysm are the most common. Congenital diseases of the aorta include supravalvular aortic stenosis, coarctation, arch interruption, aneurysm (sinus of Valsalva), and aortic rings. These conditions alter the aorta mainly by narrowing and less frequently by rupturing.

General Considerations

Normal Aorta

The aorta is divided into several anatomic segments: ascending, arch, descending (thoracic), and abdominal portions (Fig. 1). The ascending aorta may be further divided into the sinus portion (that segment from its origin to about the level of the coronary artery ostia) and the tubular portion (that segment beyond the origin of the coronary ostia). A prominent ridge separating these two areas is termed the sinotubular junction. The normal aorta consists of elastic fibers, collagen fibers, smooth muscle cells, and a mucoid ground substance. Each of these normal components change with age.³

The elastic fibers are arranged in parallel layers, are more concentrated in the ascending aorta (56 layers in ascending aorta, 28 layers in descending aorta)⁴ and fragment with advanced age, at which stage the smooth muscle cells decrease in number while the number of collagen fibers and the amount of mucoid ground substance increase. The results of these aging changes are weakening of the wall and generalized dilation (both transverse and longitudinal). This aging dilation process has been termed aortic ectasia.

Acquired Diseases of the Aorta

Atherosclerosis: The most common disease of the aorta is atherosclerosis. This lesion is characterized by involvement of the intima by deposits of amorphous lipoid material, cholesterol clefts, fibrosis, and, later, calcific deposits. When present, the lesions are more extensive in the abdominal portion of the aorta than in any other segment. In the abdominal portion, atherosclerotic plaques are confluent, whereas in the absence of aortitis the plaques in the thoracic aorta tend to be focally distributed.⁵ As the atherosclerosis plaques become complicated (extracellular lipid, fibrosis, calcific deposits), the process may cause atrophy of the underlying media. The effect of the atherosclerotic process upon the medial is purely a secondary phenomenon.

Of all portions of the aorta, the segment least prone to atherosclerosis (in the absence of aortitis or disease of the media) is the ascending aorta. Some patients have atherosclerotic

Address for reprints:

Bruce F. Waller, M.D.
Suite 400
8333 Naab Road
Indianapolis, IN 46260, USA

Received: March 3, 1997

Accepted: May 6, 1997

FIG. 1 Diagram showing segmental divisions of aorta. From Ref. No. 1 with permission.

FIG. 2 Diagram showing distribution of aortic dissection. From Ref. No. 1 with permission.

plaque diffusely covering the intimal surfaces of the abdominal, descending, and arch of the aorta, yet have no atherosclerotic plaque in the ascending aorta. In contrast, a subgroup of patients with homozygous and heterozygous type II hyperlipoproteinemia have more atherosclerosis in the ascending than in the abdominal aorta.⁶

Two other conditions may be associated with extensive atherosclerotic plaque in the ascending aorta. One is in patients with diabetes mellitus and the other is aortitis (e.g., syphilis). In diabetes mellitus, the atherosclerotic process in the ascending aorta is usually less severe than that in the abdominal aorta. In aortitis, the underlying aortic media and adventitia are abnormal, and the extensive atherosclerotic intimal process appears to be a secondary phenomenon.

Consequences of severe atherosclerosis of the aorta are luminal dilation (aneurysm), luminal obstruction, and distal emboli. Luminal dilation (saccular, fusiform aneurysms) may affect the arch, descending, and abdominal aorta. Luminal obstruction generally affects the abdominal portion of the aorta, although the ostium of any artery arising from any segment of aorta may be narrowed by atherosclerotic plaques (coronary, cerebral, mesenteric, renal, and iliac arteries). Fragments of friable atherosclerotic plaques and/or thrombus may embolize to any of the arteries arising from the aorta, although the arteries arising from the abdominal aorta are most susceptible to this complication.

When aortic obstruction is the consequence of atherosclerosis (with or without superimposed thrombus), the site of obstruction is the abdominal aorta. Why the severely atherosclerotic process involving abdominal aorta results in aneurysm formation in some patients and luminal obstruction without aneurysm in others is unknown.^{1,2} When severe atherosclerosis with superimposed thrombus causes abdominal aortic obstruction, the abdominal aorta is nondilated and the wall of involved aorta is severely calcified.² The respective temporal

occurrence of dilation and calcification may be a factor in development of aneurysm versus luminal occlusion.² If atherosclerotic plaque is associated with early and heavy calcific deposits, the abdominal aorta becomes rigid and does not have an opportunity to dilate (i.e., luminal obstruction results). If, on the other hand, the atherosclerotic process weakens the media and is associated with minimal calcific deposits, luminal dilation occurs (i.e., aneurysm formation).

Aortic dissection (Figs. 2–7): Aortic dissection, recognized at necropsy more than 200 years ago, has been known by various names: dissection, dissecting aneurysm, dissecting hematoma, intramural hematoma, false channel, and false channel hematoma. The process is simply characterized by a separation of the media. All dissections are characterized by a separation of medial layer in a course parallel to that of the flow of blood. In addition to the medial longitudinal separation, a transverse intimal and medial tear is identified in more than 95% of patients.⁷ In about 60% of patients, the tear

FIG. 3 Diagram of ascending aortic dissection extending to descending thoracic aorta showing entry and reentry side. From Ref. No. 1 with permission.

FIG. 4 Acute descending aortic (AA) dissection (D) resulting from automobile collision in which the patient suffered a steering wheel injury similar to the diagram in Figure 3. The false channel (FC) is filled with hematoma. From Ref. No. 1 with permission.

marking the beginning of the dissection (entrance tear, primary tear) is located in the ascending aorta (Figs. 2, 3) (Table I), usually about 2 cm cephalad to the sinotubular junction.^{1,7-13} The entrance tear is in the arch of the aorta in about 10%, in the descending aorta in about 25% (Figs. 4-6) and, rarely (in about 3%), in the abdominal aorta. The entrance tear, in general, involves about one-half of the aortic circumference (Fig.

FIG. 6 Diagram illustrating another mechanism for stopping an acute aortic dissection: branching arteries. The proximal dissection stops at the renal arteries. In addition, distal medial damage from a previous fusiform aortic aneurysm may have helped prevent distal progression of the proximal dissection. From Ref. No. 1 with permission.

FIG. 5 Diagram illustrating how disease of the media (scarring) from atherosclerotic plaque helps stop a progressing medial dissection. (Modified from Roberts WC: Pathology of arterial aneurysms. In *Aneurysms: Diagnosis and Treatment* (Eds. Bergan JJ, Yao JST), p. 17. Orlando: Grune & Stratton, Inc., 1982. From Ref. No. 1 with permission.

3), occasionally may involve less than one-quarter or, rarely, the entire aortic circumference (Fig. 7). The intramedial hematoma tends to progress distally from the site of entrance tear for varying distances depending on medial scarring and/or branch points (Fig. 2). The intramedial hematoma may extend proximally, but usually does so only to a limited extent.⁵ Frequently, the medial separation extends into the media of arterial branches (Figs. 3, 4). In contrast to the primary tear, a reentry tear is identified far less frequently—in about 10% of patients collected by Hirst *et al.*⁷ (Table II). In one-half of the patients, the reentry site is within the aorta (most commonly the abdominal aorta); in the remaining patients, the site of reentry is in a peripheral artery (most commonly the iliac artery) (Fig. 3).

The location of the false channel (dissecting hematoma) is in the outer half of the aortic media.¹⁵ As a result, the outer wall of the false channel is very thick in contrast to the inner wall (partition between true and false channels) of the false channel, which is composed of media and intima. The thinness of the outer wall of the false channel is the anatomic explanation for the high frequency of rupture and also explains why rupture outside the aorta (into adjacent spaces) is more common than rupture of the false channel into the true channel.¹⁵ Barsky and Rosen¹⁶ observed aortic wall necrosis in 21 (62%) of 34 necropsy patients with aortic dissection. These authors noted that among patients with aortic dissection surviving as long as 48 h, necrosis of the aortic wall may occur, further weakening the aortic wall and promoting rupture. Although the entrance tears are most frequently located in the ascending aorta, the descending aorta is the most common portion of the aorta to be involved in the dissection process. This is because

FIG. 7 Diagram shows circumferential aortic dissection with intussusception of partition between true and false channels causing sudden death. From Ref. No. 1 with permission.

most ascending aortic dissections extend to the descending aorta and few descending aortic dissections extend retrograde to the ascending aorta (Figs. 2–4).¹⁵

Roberts has addressed the question of why and where aortic dissections stop.¹⁵ The cause of interruption of the longitudinal dissection appears to be medial scarring (Fig. 5). The most

common cause of medial interruption is extensive atherosclerotic plaque that has secondarily caused underlying medial scarring, atrophy, or both.¹⁵ Extensive atherosclerosis of the

TABLE I Aortic dissection: Sites of primary tear in aorta

Portion of aorta	Number (%)
Ascending	244 (62)
Arch	37 (9)
Isthmus	62 (16)
Descending thoracic	41 (10)
Abdominal aorta—partial	10 (3)
Totals	394 (100)

From Ref. No. 7 with permission.

TABLE II Aortic dissection: Sites of secondary or reentry tear

Artery	Number (%)
Aorta	33 (50)
Ascending or arch, 2	
Isthmus and descending thoracic, 14	
Abdominal, 17	
Peripheral	33 (50)
Iliac, 22	
Carotid, 1	
Subclavian, 1	
Not specified, 9	
Totals	66 (100)

From Ref. No. 7 with permission.

abdominal portion of the aorta (with or without aneurysm) may also prevent further longitudinal dissection (Fig. 6).¹⁵ Other causes of scarred media include aortitis and inflammatory conditions such as syphilis. In cardiovascular syphilis, focal areas of medial and adventitial scarring may interrupt longitudinal dissection. Another cause of interruption of a dissection involving the ascending and/or arch portions of the aorta is aortic isthmus coarctation. Because aortic coarctation represents an invagination of the media,¹⁷ progression of a longitudinal dissection starting in the proximal aorta cannot extend through the coarctation site (or previously resected coarctation site). Aortic dissection in patients with aortic coarctation most commonly involves only the aorta proximal to the coarctation site, occasionally only distal to the coarctation site, but never does it involve both areas.^{1, 15, 17-21} Of 200 necropsy patients with coarctation of aorta collected by Abbott,¹⁷ 38 (19%) had aortic rupture occurring proximal to the coarctation site, and in 5 patients, rupture occurred distal to the coarctation site. Thus, without extensive aortic atherosclerosis, previous medial inflammation, or aortic coarctation, the entire aorta distal to the entrance tear usually dissects.¹⁵ We recently examined a patient with sudden death who, at necropsy, had aortic dissection limited to the ascending aorta (Fig. 7). Dissection started in the proximal ascending aorta, extended to the beginning of the aortic arch, and then stopped because of heavy calcified plaque. Progressive proximal dissection expanded the entrance tear to involve the entire aortic circumference. With separation of the intimal flap, intussusception of the partition between true and false channels caused near-total occlusion of the distal aortic lumen (Fig. 7).

References

- Tejada E, Waller BF: Non-neoplastic diseases of the aorta and pulmonary trunk. In *Pathology of the Heart and Great Vessels*, Chapter 11 (Ed. Waller BF), p. 367-427. New York: Churchill Livingstone, 1988
- Roberts WC: The aorta: Its acquired diseases and their consequences as viewed from a morphologic perspective. In *The Aorta* (Eds. Lindsay J, Hurst JW), p. 51. Orlando: Grune & Stratton, 1979
- Schlatmann TJM, Becker AE: Histologic changes in the normal aging aorta. *Am J Cardiol* 1977;39:13-21
- Wolinsky H, Glagov S: A lamellar unit of aortic medial structure and function in mammals. *Circ Res* 1967;20:99-107
- Edwards JE: Manifestations of acquired and congenital diseases of the aorta. *Curr Probl Cardiol* 1979;3(1):1-51
- Roberts WC, Ferrans VJ, Levy RI, Fredrickson DS: Cardiovascular pathology in hyperlipoproteinemia. Anatomic observations in 42 necropsy patients with normal or abnormal serum lipoprotein patterns. *Am J Cardiol* 1973;31:557-565
- Hirst AE Jr, Johns VJ Jr, Kime SW Jr: Dissecting aneurysm of the aorta: A review of 505 cases. *Medicine* 1958;37:217-228
- Erb BD, Tullis IF: Dissecting aneurysm of the aorta. The clinical features of thirty autopsied cases. *Circulation* 1960;22:315-320
- Lindsay J Jr, Hurst JW: Clinical features and prognosis in dissecting aneurysm of the aorta: A re-appraisal. *Circulation* 1967;35:880-888
- Anagnostopoulos CE, Prabhakar MJS, Kytte CF: Aortic dissections and dissecting aneurysms. *Am J Cardiol* 1972;30:263-269
- Gore I, Hirst AE Jr: Dissecting aneurysm of the aorta. *Cardiovasc Clin* 1973;2:239-247
- Talbot S: Clinical features and prognosis of dissecting aneurysm and ruptured saccular aneurysm. *Chest* 1974;66:252-260
- Slater EE, DeSanctis RW: The clinical recognition of dissecting aortic aneurysm. *Am J Med* 1976;60:625-631
- Murray CA, Edwards JE: Spontaneous laceration of the ascending aorta. *Circulation* 1973;47:848-852
- Roberts WC: Aortic dissection: Anatomy, consequences, and causes. *Am Heart J* 1981;101:195-207
- Barsky SH, Rosen S: Aortic infarction following dissecting aortic aneurysm. *Circulation* 1978;58:876-882
- Abbott ME: Coarctation of the aorta of the adult type II. A statistical study and historical retrospect of 200 recorded cases with autopsy of stenosis or obliteration of the descending arch in subjects above the age of two years. *Am Heart J* 1928;3:392-401
- Edwards JE, Christensen NA, Clagett OT, McDonald JR: Pathologic considerations in coarctation of the aorta. *Proc Staff Meet Mayo Clin* 1948;23:324-328
- Reifenstein GH, Levine SA, Gross RE: Coarctation of the aorta. A review of 104 autopsied cases of "adult type," 2 years of age or older. *Am Heart J* 1947;33:146-150
- McCombs HL, Crocker DW: Dissecting aneurysm distal to coarctation of the aorta, with long survival. *Am Heart J* 1967;74:675-681
- Edwards JE: Aneurysms of the thoracic aorta complicating coarctation. *Circulation* 1973;48:195-201