4 Pathology of the Aorta: Inflammatory and Noninflammatory Conditions Predisposing to Aneurysm Formation, Dissection, and Rupture

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Introduction

The aorta is an elastic artery with a caliber that expands with systole and recoils during diastole. In adults, it extends an average of 490 cm in length from thoracic initiation to pelvic bifurcation, and it is anatomically divided into three layers, i.e. the tunica intima, tunica media, and tunica adventitia (Fig. 4.1) [1]. The tunica intima is the layer closest to the aortic lumen and is composed of extracellular matrix proteins and a few multipotent stromal cells [1, 2]. It includes an endothelial cell layer and extends to the internal elastic lamina (IEL). The IEL delimits the tunica intima from tunica media and consists of elastic fibers that form a barrier between large molecules within the circulating blood (e.g. cholesterol) and the underlying layers of the aortic wall. In newborns, the intimal layer is quite thin with the endothelial cells closely approximated to the IEL; with "wear-and-tear" aging, intimal thickness increases due to deposition of extracellular matrix proteins [3]. The tunica media extends to the external elastic lamina (EEL) and is the thickest layer of the aorta. Its components are concentrically arranged into lamellar units with each lamellar unit being composed of a layer of elastic fibers with subjacent smooth muscle cells and some embedded extracellular matrix including collagen fibers and ground substance [1, 2]. In newborns, the number of stacked lamellar units is only about 35; however, by adulthood, this has generally increased to 50–60 lamellar units [3]. The tunica adventitia is the outermost layer of the aortic wall and is composed of connective tissue, adipocytes, lymphatic channels, and the vasa vasorum. Given the width of the aortic wall, a specialized vascular system is needed to supply oxygen and other nutrients to the parts of the aortic wall furthest from the blood flowing within the aortic lumen; thus, the vasa vasorum supplies oxygen and nutrients to the outer third of the tunica media. Disruption to the normal layers of the aortic wall due to a variety of diseases can result in significant morbidity and mortality. Thus, familiarity with clinicopathologic features supportive of particular disease processes is advised.

Overview of Aortic Disease

The recent consensus statement on surgical pathology of the aorta from the Society for Cardiovascular Pathology recommends that aortic diseases detected in surgical pathology specimens be categorized as either inflammatory or noninflammatory processes and offers a standardized approach to the processing of specimens, disease grading, and disease nomenclature [3, 4]. It is anticipated that such standardization will result in enhanced understanding of aortic diseases and improved patient care. Either inflammatory or noninflammatory aortic disease may ultimately result in serious sequelae such as aortic aneurysm formation, acute aortic dissection, or aortic rupture. A brief overview of the pathogenesis and epidemiologic features of aortic aneurysms and aortic dissections is followed by a review of the distinct inflammatory and noninflammatory disease processes that may lead to such phenomena.

Aortic Aneurysm

Pathologically, a true aortic aneurysm is defined as a localized dilatation involving the entire thickness of the wall; it may be congenital or acquired [5]. A false or pseudoaneurysm, in contrast, is defined as a ruptured arterial wall in which blood is confined by surrounding tissues forming an extravascular hematoma [5]. In the United States, true thoracic and abdominal aortic aneurysms represent the 15th leading cause of death in people greater than age 55 and are the 19th overall leading cause of death [6].

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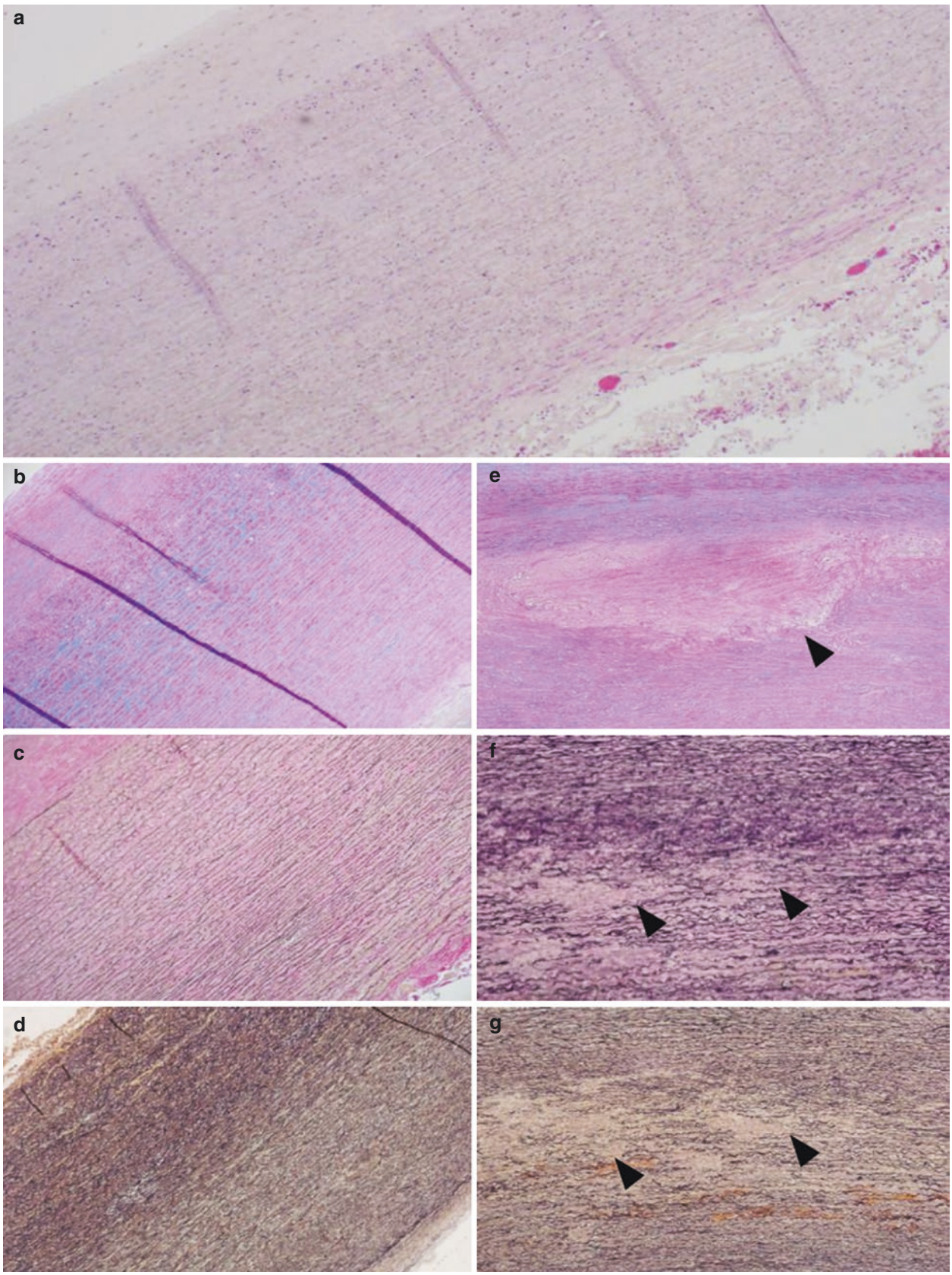

Fig. 4.1 (**a**) Normal layers of the aortic wall with the tunica intima, tunica media, and tunica adventitia. (**b**) There is no medial degeneration present (Alcian Blue/PAS stain). (**c**) The normal aorta contains elastic fibers in an organized pattern (Elastic stain). (**d**) Organized elastic fibers without any degenerative changes (Movat's pentachrome stain). (**e**) Medial degeneration within the aortic wall (arrowhead). (**f**) Elastic fiber fragmentation and loss (arrowheads). (**g**) Increased extracellular matrix material (arrowheads)

Thoracic Aortic Aneurysm

More than 95% of thoracic aortic aneurysms (TAAs) are asymptomatic; the prevalence of clinically silent TAAs is estimated to fall between 0.16% and 0.34% [6]. TAAs are more common in men compared to women [6, 7]. The TAA distribution in 60% occur in the root or ascending aorta, 10% in the arch, 40% in the descending aorta, and 10% in the thoracoabdominal aorta (Fig. 4.2). The incidence of TAAs appears to be increasing [6, 7]. There seems to be a genetic component to TAAs with 21% of patients with a TAA having a family member with some sort of aneurysm. Genetic syndromes such as Marfan syndrome, Ehlers-Danlos syndrome, Loeys-Dietz syndrome, and Turner syndrome can have TAAs as part of their clinical manifestations of the genetic abnormality [6]. Other risk factors for development of thoracic aortic aneurysm include aging, systemic hypertension, and bicuspid aortic valve [7].

Histologically, medial degeneration is commonly seen in thoracic aortic aneurysm resections. Medial degeneration leads to a weakened aortic wall which predisposes to dilatation and eventual aneurysm formation. Mucoid extracellular matrix accumulation, loss of smooth muscle cells with disarray, elastic fiber degeneration, and disorderly arrangement are some features of medial degeneration [3, 7]. In patients with Marfan syndrome, mutations in fibrillin-1 are found. Fibrillin-1 is important to the construction of microfibrils in the extracellular matrix. Medial degeneration is often significant in patients with Marfan syndrome [7].

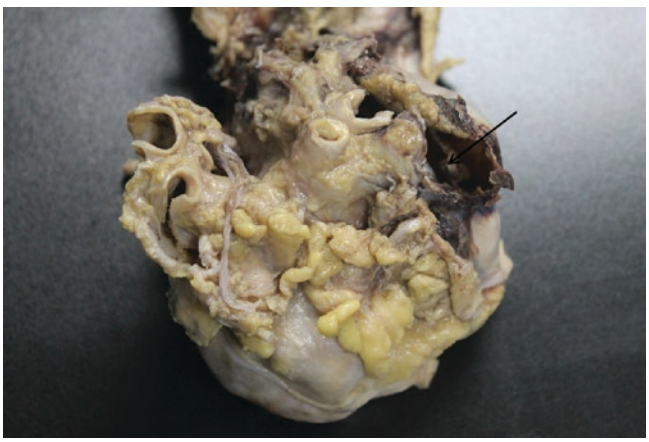

Fig. 4.2 Thoracic Aortic Aneurysm Rupture with hemorrhage (arrow demonstrating aortic arch rupture). Brachiocephalic Artery, Left Common Carotid Artery, and Left Subclavian Artery are adjacent to area of rupture

Abdominal Aortic Aneurysm

Abdominal aortic aneurysms (AAAs) are more common in occurrence when compared to TAAs; up to 80% of aortic aneurysms occur below the renal arteries [7]. Most are due to atherosclerosis, but other causes such as inflammatory aneurysm, tuberculous aneurysm, medial degeneration, or infectious aneurysm are possible [7]. Risk factors for AAAs include smoking, increasing age, greater height, coronary artery disease, atherosclerosis, hypercholesterolemia, and hypertension [7, 8]. Like thoracic aortic aneurysms, genetics also can play a role in AAAs with 12–19% of those patients having an abdominal aneurysm repair being related to a first degree relative with an AAA [8]. In addition, patients with Ehlers-Danlos type IV who have a defect in their type III collagen synthesis are at an increased risk of AAA [8].

Atherosclerotic plaques play a role in development of AAAs. Significant inflammatory infiltrates including numerous macrophages and lymphocytes are common in AAAs which may contribute to extracellular matrix degeneration. Histologically, elastic fiber loss, medial smooth muscle cell degeneration, and fibrosis can be seen. Another common finding is thrombus formation within the aortic lumen associated with the aneurysm. Enzymes such as collagenase and elastase may be upregulated contributing to aneurysmal dilatation [7].

Aortic Dissection

Approximately 3/100, 000 persons/year develop an aortic dissection with a slight male predominance. Risk factors for an aortic dissection include age, inherited genetic connective tissue diseases such as Marfan syndrome and Ehlers-Danlos syndrome, hypertension, aortic valvular disease such as bicuspid aortic valve, trauma, prior aortic operations, and cocaine abuse. The most common symptoms are back, abdominal, or chest pain [9]. An aortic dissection occurs when a tear develops in the intimal layer allowing blood to enter into the tunica media layer creating a true lumen and a false lumen (Figs. 4.3, 4.4, and 4.5). In cases related to an intimal tear, 60% occur in the ascending aorta, 25% in the descending aorta, and 10% in the arch and abdominal aorta [9]. Other causes of aortic dissection could be bleeding from the vasa vasorum in the tunica adventitia into a weakened tunica media layer.

Aortic dissections are often classified into two types: Stanford Type A which occurs in the ascending aorta and Stanford Type B which originates in the descending aorta [9]. Approximately 60% of patients develop Type A dissections with the incidence peaking between 50–60 years of age [9]. In patients with inherited connective tissue diseases, a common histological finding is medial degeneration which is

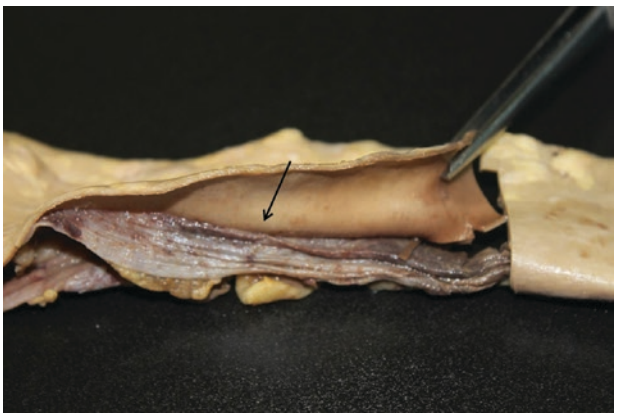

Fig. 4.3 Aortic dissection with true lumen and false lumen (arrow indicating false lumen)

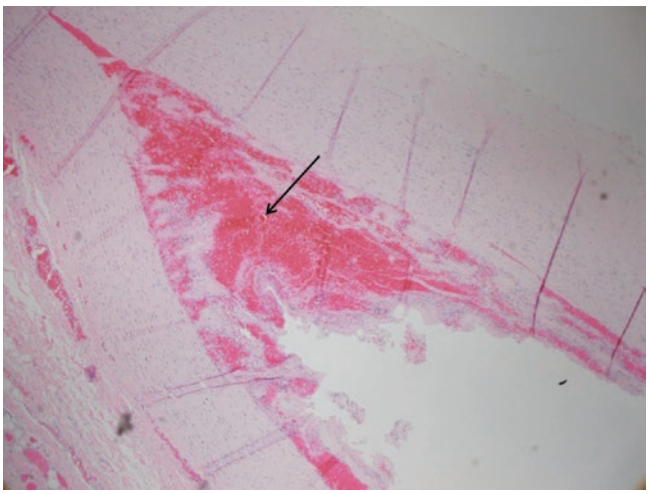

Fig. 4.4 Aortic dissection with hemorrhage (arrow) present within the separating tunica media

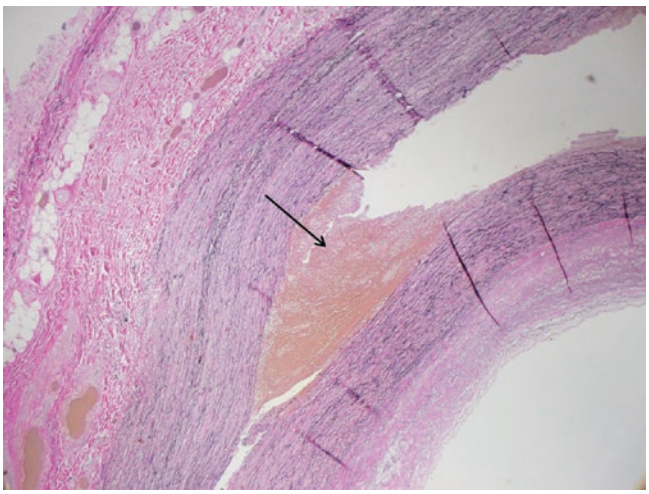

Fig. 4.5 Aortic dissection with elastic fibers stained with an elastic stain showing dividing layers of tunica media as hemorrhage accumulates within the tunica media

characterized by loss of elastic fibers in the tunica media, decrease in smooth muscle cells, and an increase in proteoglycans, glycosylated proteins [3, 9, 10]. In patients without connective tissue diseases, medial degeneration may be milder or less common. Electron microscopy of the layers of the aortic wall has shown in normal aortic walls elastic fibers are organized longitudinally but also with connections between the longitudinal elastic fibers. In patients without a connective tissue disease, it is possible that loss of connection between longitudinal elastic fibers is a mechanism in the pathogenesis of aortic dissection [10].

Noninflammatory Conditions of the Aorta

Degenerative changes of the aortic wall can result from a variety of noninflammatory conditions such as aging, hypertension, genetic syndromes, and congenital anomalies. Histologically, they may show medial degeneration. Medial degeneration includes mucoid extracellular matrix accumulation, elastic fiber fragmentation, thinning, and disorganization, smooth muscle cell nuclei loss and disorganization, laminar medial collapse, and medial fibrosis (Figs. 4.6 and 4.7) [1, 3]. Each of these subcategories are components of medial degeneration and can be graded as mild, moderate, and severe to contribute to the overall grading of medial degeneration as mild, moderate, and severe [3]. Aging, genetic syndromes, congenital diseases, hypertension, cocaine, intense physical exertion, and pregnancy can be associated with degenerative changes within the tunica media [1, 3]. Attempting to streamline the pathologic terminology used to describe abnormalities within the aortic wall may improve understanding of the common histologic appearance of the various causes of aortic pathology [3].

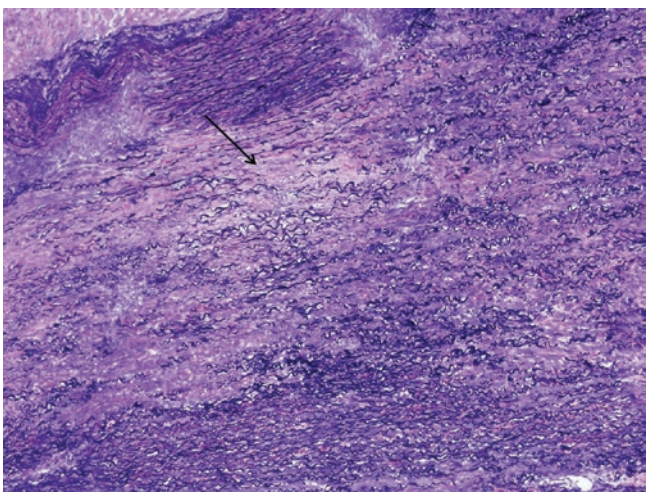

Fig. 4.6 Elastic fiber fragmentation as seen in aging and noninflammatory conditions of the aorta previously described

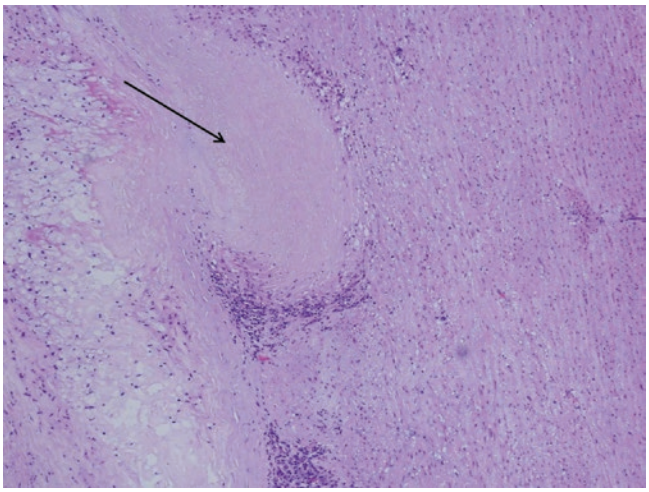

Fig. 4.7 Medial degeneration as previously described in noninflammatory conditions of the aorta

Aortic Disease in the Elderly

As individuals age, the aorta becomes less pliable and enlarges placing them at risk for aneurysm and dissection. Within the thoracic aorta, the diameter changes as people age. However, in the abdominal aorta, the most significant change is an increase in stiffness [1]. Elastin decreases and becomes more fragmented. In addition, there is intimal proliferation and increased disorganized collagen. Smooth muscle cells become less prevalent. Each of these changes presents a challenge for the aorta to repair itself when injury occurs [1].

Aortic Disease in the Young

Genetic syndromes such as Marfan syndrome, Loeys-Dietz syndrome, Turner syndrome, Ehlers-Danlos syndrome, Familial thoracic aortic aneurysm and dissection, Arterial tortuosity syndrome, Shprintzen-Goldberg, and autosomal dominant polycystic kidney disease can demonstrate elastic fiber fragmentation and loss [1, 3]. Marfan syndrome (MFS) results from a genetic abnormality in the fibrillin-1 gene which predisposes 68–80% of individuals to aortic root dilatation by age 19 [11]. Individuals with this syndrome may have pectus excavatum, arachnodactyly, tall stature, lens ectopia, and mitral valve prolapse [1, 11]. At autopsy, annuloaortic ectasia where both the aortic annulus and ascending aorta are enlarged with a flask-like shape may be seen. Histologically, elastic fiber fragmentation and medial degeneration are observed [1, 3, 11].

In Turner syndrome, a sex aneuploidy syndrome where individuals have a single X chromosome, 1.5% of individuals have aortic dilation along with other cardiac abnormalities such as bicuspid aortic valve and coarctation [11]. Other manifestations of Turner syndrome include webbed-neck, short stature, and lymphedema [1]. By contrast, individuals with Ehlers-Danlos syndrome type IV have an abnormality in their collagen, type III, alpha-1 (COL3A) gene which encodes type III 1 collagen found in skin, vessel walls, and hollow organs [11]. Individuals may have thin skin, a thin pinched nose, thin lips, prominent ears, and be prone to bruising [1]. Rupture and dissection tend to be more common than aortic aneurysm formation.

In individuals with autosomal dominant polycystic kidney disease, a mutation found in polycystic kidney disease (PKD) 1 gene, can result in saccular intracranial aneurysms and at times abdominal aortic aneurysm or thoracic aortic aneurysm rupture [1, 11]. Other manifestations include renal cysts and renal failure [11]. Aneurysm rupture could be related to hypertension within these individuals. About 20% of patients being seen for thoracic aneurysm repair have clustering of thoracic aneurysms in their family. Inheritance is usually autosomal dominant and can be caused by a number of different genetic mutations [1, 11].

Loeys-Dietz syndrome, a disorder of connective tissue diseases, shares some features with MFS and vascular type of Ehlers-Danlos syndrome [1, 11]. Individuals may have hypertelorism, changes to their uvula, cleft palate, craniosynostosis, and be at risk for visceral rupture as well as easy bruising [1].

Aortic tortuosity syndrome can occur due to a mutation in solute carrier family 2 member 10 gene which encodes a glucose transporter (GLUT10) [11]. Medial degeneration, specifically, elastic fiber fragmentation may be observed histologically [1].

Congenital

Bicuspid aortic valve affects approximately 2% of the population and is the most common congenital heart abnormality. The cause of bicuspid aortic valve is mostly unknown. About 50% of young men with bicuspid aortic valve have aortic dimensions fitting with an aneurysm and 5% develop an aortic dissection [11]. NOTCH-1 mutations can be seen in those with bicuspid aortic valve [1]. Individuals may develop an aneurysm of the aortic root which is associated with a sixfold increased dissection risk [1]. Medial degeneration within these aortas can range from mild to severe [1]. Other congenital anomalies, such as coarctation of the aorta and narrowing of the aorta distal to the left subclavian artery, can be associated with bicuspid aortic valve and rarely aortic dissection. Histologically, mucoid extracellular matrix accumulation and elastic fiber fragmentation may be observed. Finally, most patients with tetralogy of Fallot undergo repair

but can develop aortic root dilatation as they age. Pathology often demonstrates medial degeneration with lack of lamellar units, elastic fiber fragmentation, mucoid extracellular matrix accumulation, and medial fibrosis [1].

Other Noninflammatory Causes of Aortic Aneurysm and Dissection

Systemic hypertension is one of the most important risk factors for medial degeneration and development of aortic aneurysm and dissection [1, 3]. In a Mayo Clinic study of 513 patients requiring resection of a thoracic aortic aneurysm, 54.4% had a history of hypertension [12]. Hypertension may accelerate the aortic aging process through altered hemodynamic forces which greatly affect the aortic media [1, 12].

Cocaine abuse is associated with multiple cardiovascular abnormalities including aortic dissection. Cocaine causes release of catecholamines which stimulate alpha- and beta-adrenergic receptors which can lead to vasoconstriction and arterial spasm. Chronically, cocaine leads to increased diastolic aortic diameter, loss of aortic elasticity, and increased aortic stiffness [11]. Histologically, resected aortas can demonstrate mucoid extracellular matrix accumulation [1]. When combined with uncontrolled hypertension and smoking, these individuals can be at risk for aortic dissection [11].

Aortic dissection can be seen in weightlifting and severe physical exertion. Typically, patients have preexisting moderately enlarged aortas. The cause of the aortic dissection in this situation could be related to a rapid and extreme elevation in blood pressure during significant exertion against an already dilated aorta [11]. Intense physical exertion can elevate blood pressure over 300 mmHg which increases wall stress [3]. Microscopically, mucoid extracellular matrix accumulation has been seen. The stress on the aortic wall may lead to aortic rupture [3].

Pregnancy can also affect the aortic wall. In women with genetic predisposition to an aortic dissection, such as individuals with MFS, bicuspid aortic valve, an existing aneurysm, or an aortic diameter greater than 4 cm, there is a risk for aortic dissection. The etiology is thought to be from hormonal influences leading to elastic fiber fragmentation and a decrease in aortic wall mucopolysaccharides [3].

Inflammatory Conditions of the Aorta

Inflammatory conditions of the aorta include atherosclerosis, aortitis, and periaortitis [2, 4]. Inflammation of the aorta can result in aneurysm, aortic wall rupture, acute dissection, and obstruction of the aortic lumen [4].

Atherosclerosis

Atherosclerosis is a pathologic process that may cause disease of the cerebral, coronary, mesenteric, and peripheral arteries. Men and women aged 15–34 years of age have been found to have the earliest form of atherosclerosis known as fatty streaks. Risk factors for atherosclerosis include smoking, diabetes mellitus, dyslipidemia, and hypertension. The presence of aortic atherosclerosis indicates systemic atherosclerosis is present.

The pathophysiology of atherosclerosis involves lipid disturbances, platelet activation, thrombosis, endothelial abnormalities, inflammatory responses, oxidative stress, smooth muscle cell proliferation, vascular remodeling, and genetic characteristics.

Classification of degree of severity of atherosclerosis within the aorta appears to differ from classification schemes used in the coronary arteries [4]. Most classification models are based on changes within the tunica intima, but within the aorta the consequences of atherosclerosis are mainly based on changes within the tunica media [4]. Within the aorta, atherosclerosis is categorized as mild, moderate, or severe [4]. No significant atherosclerosis consists of normal or fatty streaks with intimal thickening or hyperplasia and foam cells and lymphocytes seen histologically. Mild atherosclerosis consists grossly of raised plaques and extracellular lipid deposition without fibrosis microscopically. Moderate atherosclerosis consists of raised or confluent plaques which has extracellular lipid deposition and fibrosis as well as destruction of the media up to 1/3 of thickness. Severe atherosclerosis also consists of raised or confluent plaques on gross examination as well as extracellular lipid deposition with fibrosis, but media destruction constitutes more than 1/3 of the thickness. Importantly, pathologists must also comment if plaque disruption, luminal thrombus, or if the plaque is calcified. Noting the presence of thrombus is important for possible embolic events [4].

Atherosclerosis begins as a fatty streak (Fig. 4.8). The fatty streak develops when foam cells (macrophages filled with lipids) accumulate in the intima of an artery. Smooth muscle cells are also present in the intimal layer of arteries and begin to collect. Smooth muscles cells may undergo apoptosis as they increase in the intimal layer which attracts additional macrophages with microvesicles capable of calcifying. This process may facilitate transition of the fatty streak to an atherosclerotic plaque [13].

Once an atherosclerotic plaque develops, there is risk of plaque ulceration or thrombosis (Fig. 4.9). As plaques develop, an extracellular lipid core accumulates covered by a fibrous cap which contains smooth muscle cells and macrophages forming an atheroma (Fig. 4.10) [13, 14]. In plaques with the volume composed predominantly of extracellular lipid and a fibrous cap made largely of macrophages instead

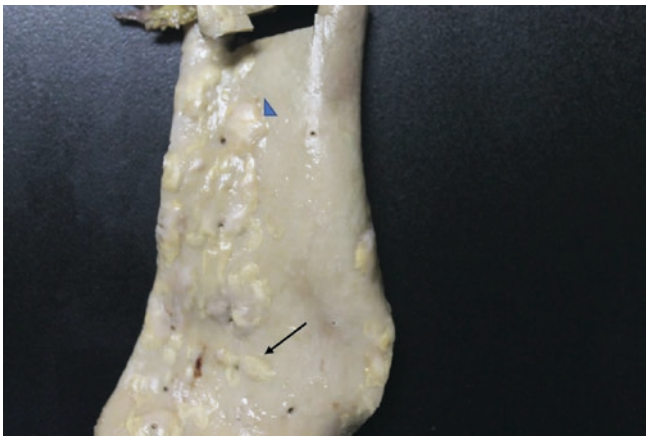

Fig. 4.8 Mild atherosclerosis with both fatty streaks (arrow) and atherosclerotic plaques (arrow head) present

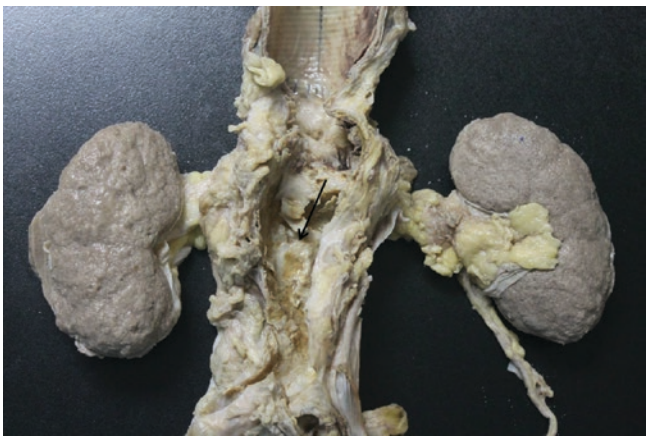

Fig. 4.9 Severe atherosclerosis with ulceration of atherosclerotic plaques (arrow) and shrunken kidneys bilaterally. Graft present superior to area of atherosclerotic plaque ulceration

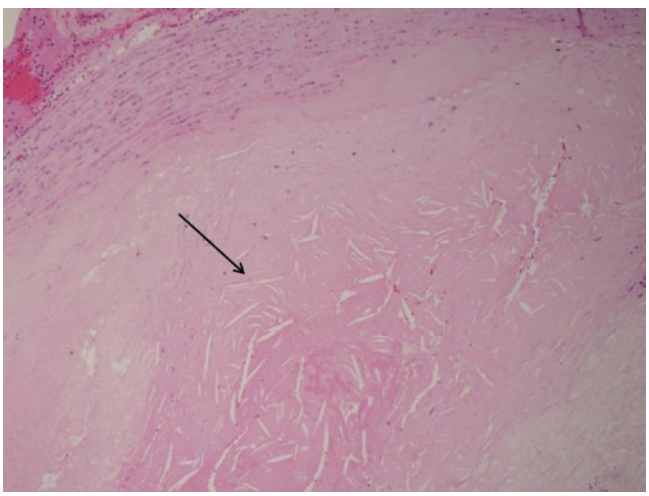

Fig. 4.10 Atheroma within the aortic wall (arrow indicating atheroma and disruption of tunica media)

of smooth muscle cells, thrombosis and ulceration can be seen [14]. Lymphocytes can be observed in the intima, media, and/or adventitia which are mainly CD3+ T-cells, but CD20+ B-cells and plasma cells can also be seen [4]. Disrupted plaques may contain surface thrombus, or if the plaque has existed for a long time, organizing thrombus may be observed. In addition, at the site of disruption, neutrophils can be identified. Cholesterol clefts can cause a giant cell reaction [4].

Some moderate to severe cases of atherosclerosis can have an unusually intense inflammatory infiltrate. Neutrophils can concentrate around the disrupted plaque. Differentiating from bacterial infection can be done by noting absence of necrosis except for in the lipid atherosclerotic core and absence of microorganisms. Gram stain, Grocott-Gomori methenamine silver stain, Steiner, or Warthin-Starry stain may be helpful in identifying infectious causes [4].

Severe atherosclerosis can be associated with an inflammatory atherosclerotic aneurysm especially within the abdominal aorta. Histologically, a lymphocytic, plasma cell, and possibly eosinophilic inflammatory infiltrate is observed within the aortic adventitia along with a severe atherosclerotic lesion. Lack of granulomas, IgG4+ plasma cells, purulent inflammation, or significant necrosis helps distinguish inflammatory atherosclerotic aneurysm from periaortitis.

Aortitis

Ascending aortitis can be a cause of aortic aneurysm. Inflammation of the adventitia and media with giant cells can occur in diseases such as Takayasu arteritis, giant cell arteritis, and isolated arteritis. Takayasu arteritis typically affects individuals between 10 and 30 years of age with a female predominance [2]. Grossly, the aorta appears thickened and rigid due to transmural fibrosis. Areas of narrowing alternating with dilatations can be observed. Histologically, inflammation can be seen in the vasa vasorum initially consisting mainly of T cells. Eventually, inflammation extends to the media and adjacent adventitia. The tunica media can have infiltrates of inflammatory cells including giant cells which lead to elastic fiber and smooth muscle cell loss. When there is destruction of the media, aneurysms can develop. Up to 45% of patients develop aneurysms most commonly in the ascending aorta and aortic arch [11].

Giant Cell Arteritis

Giant cell arteritis is the most common systemic vasculitis to affect the aorta. Patients who develop giant cell aortitis typically lack systemic symptoms usually associated with giant cell arteritis such as unilateral headache, jaw claudication,

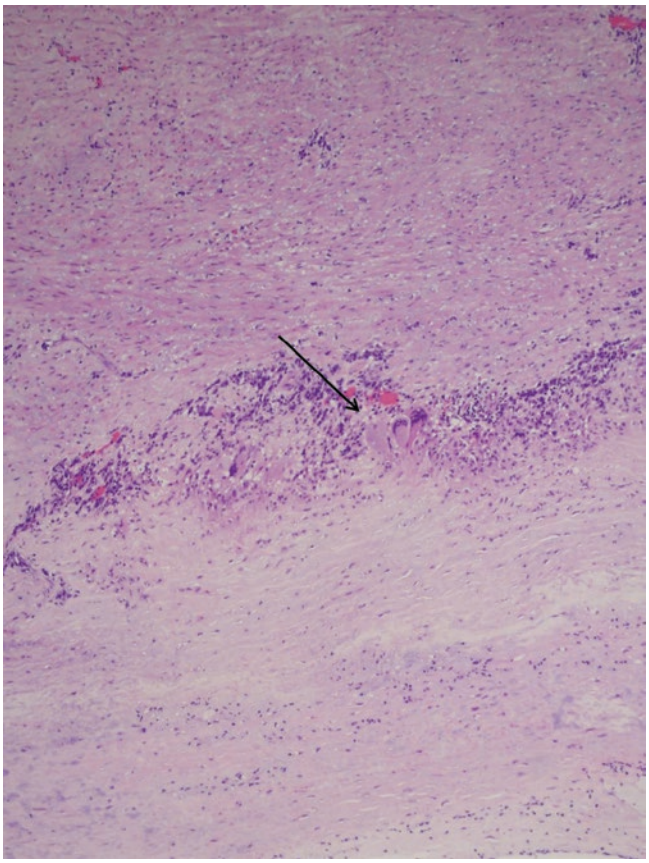

Fig. 4.11 Giant cell aortitis with giant cells and lymphocytic infiltrate (arrow indicating giant cell and surrounding lymphocytic infiltrate within the tunica media of the aorta)

and visual impairment [4]. Aortic intima begins to appear like tree bark due to medial damage, tissue edema, and inherent elastic properties of the vessel itself. Granulomatous inflammatory cells with multinucleated giant cells are seen in the tunica media (Fig. 4.11) [2]. The inner half of the media is predominantly affected [4]. Medial injury results in a moth eaten appearance on elastic stain of the tunica media. When the vasa vasora are involved, the media may infarct and lead to laminar medial necrosis which is seen in both inflammatory and noninflammatory aneurysms [2]. Within the adventitia, a lymphoplasmacytic infiltrate can develop [4]. Those who develop biopsy-proven giant cell arteritis involving temporal or occipital arteries are seventeen times more likely to develop a thoracic aortic aneurysm compared to their healthy age- and sex-matched individuals [2]. In one study, 4% of patients with giant cell arteritis on biopsy developed an aortic dissection.

Takayasu Arteritis

For individuals under the age of 50, particularly adolescent girls and women in their twenties and thirties, Takayasu arteritis is the systemic vasculitis typically affecting the aorta. The disease targets large elastic arteries. Symptoms can be mild such as impalpable pulses or more severe such as subclavian steal syndrome and cerebrovascular events. Geographically, countries such as Japan, Southeast Asia, India, and Mexico have more disease prevalence. Grossly, the aorta may dilate, develop aneurysms, or thrombosis. Inflammatory infiltrates made of granulomas, lymphocytes, plasma cells, and eosinophils can be seen. Medial necrosis with adventitial fibrosis is a common histologic pattern. Giant cell arteritis may appear similarly to Takayasu arteritis. However, Takayasu arteritis typically has a greater aortic wall thickness, and giant cell arteritis usually lacks severe adventitial scarring and inflammation predominantly affects the inner tunica media [4].

Syphilitic Aortitis

Syphilitic aortitis commonly involves the thoracic and ascending aorta with lymphoplasmacytic infiltrates seen in the media and adventitia with obliterative endarteritis in the vasa vasora at times [4]. Warthin-Starry stain can be helpful in identifying the *Treponema pallidum* microorganisms [2]. Aneurysms caused by syphilitic aortitis can erode into nearby tissue whether aortic rupture or dissection is present or not.

Pyogenic Aortitis

Pyogenic aortitis results when bacteria implant on the intimal layer of the aorta which can occur when a patient has bacteremia or endocarditis. Aortic graft placement, spread from an extravascular infection, traumatic inoculation, or embolization of bacteria to the vaso vasora can also result in pyogenic aortitis. Staphylococcus aureus, Streptococcus species, Salmonella species, Gram negative rods, and fungi have been seen in infectious aneurysms. Neutrophils can be seen within the wall of the aorta. Staining for bacteria and fungi can be done, but intraoperative cultures are preferable for diagnosis [2]. If fresh tissue or purulent material was not sent for culture from the operating room, then it is best to send fresh sterile tissue as soon as possible [4]. Additionally, fresh sterile tissue can be used to identify microorganisms by DNA amplification and sequencing. When received, the fresh sterile tissue can be frozen in order to accomplish this [4].

Aneurysm can result from all types of inflammatory aortic disease. Infectious causes usually produce noncircumferential aneurysms that may affect multiple areas of the aorta [2]. Quickly addressing the infection with debridement or an operation could lower the risk of the patient developing a dissection or rupture [2].

Surgical Pathology Processing of the Aorta

When grossing an aortic specimen, it is important when aortic tissue is received to orient, measure, and photograph the specimen. The diameter should be measured and examined for the presence of dissection, intimal tears, presence of atherosclerotic plaques, intimal thickening, evidence of prior operation, and the presence or absence of thrombus material. Decalcification of greatly calcified areas should be done. If Ehlers-Danlos syndrome type IV is suspected, the aortic media should be put in glutaraldehyde for inspection. A minimum of six full thickness sections perpendicular to the lumen should be taken. Evaluation should include a hematoxylin-eosin slide and elastic stain to evaluate the elastic fibers within the aorta. A collagen stain and an Alcian blue periodic acid Schiff stain can be helpful in identifying scarring or accumulation of collagen, proteoglycans, and mucopolysaccharides [11]. If aortitis is suspected, it is recommended to submit at least 1 section of tissue per centimeter or 12 blocks of tissue and to section the specimen perpendicular to the longitudinal axis of the aorta [4].

Conclusion

In summary, it is important for Pathologists and Surgeons to collaborate about the clinical suspicions, findings at the time of operation, identified gross abnormalities, and ultimately the microscopic findings of the aortic pathology. Both noninflammatory conditions and inflammatory conditions of the aorta may lead to aortic aneurysm, aortic dissection, and aortic wall rupture. The most complete understanding of the underlying disease process and etiology of the patient's presentation develops from understanding both the anatomy and histology of the aortic pathology and ultimately using this understanding to guide diagnosis and management of the patient's disease.

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