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Review Article

Intramural Hematoma

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ACUTE AORTIC SYNDROMES (AAS) are vascular emergencies that are associated with significant short- and long-term morbidity and mortality.[1,2] AAS includes intramural hematoma (IMH), penetrating aortic ulcer (PAU; the "A" may also be "atherosclerotic"), aortic dissection (AD), aortic rupture/transection, and an expanding/symptomatic aortic aneurysm (thoracic and/or abdominal) (Table 1).[2,3] Because AAS are defined by their acuity, an aortic aneurysm is generally not included in the group of AAS, unless it is symptomatic or has shown rapid expansion (≥ 0.5 cm/y). The incidence of AAS is estimated to be 2.6 to 3.5 cases per 100,000 person-years, the majority of which are AD (65%-75%), followed by IMH (4%-32%), and PAU ($< 10\%$).[1,4–13] Multiple lesions may coexist.[7,13]

This review focuses on IMH, which can be a particularly challenging diagnosis with potentially dire complications. Familiarity with anatomy, pathology, diagnosis, management, prognosis, and outcomes of IMH—which differ from other AAS—is the objective of this review.

Normal Aortic Anatomy

Aortic injuries are described by the layer(s) and segment(s) involved, aortic diameter and wall thickness, and associated complications. Knowledge of the normal aortic anatomy is important to understanding the pathology of IMH. The aortic wall consists of 3 layers: intima, media, and adventitia[2,7] (Fig 1). The intima (tunica intima) is the innermost layer and is in direct contact with luminal blood. It consists of a single-cell layer of endothelial cells supported by delicate connective tissue and an internal elastic lamina. The medial layer (tunica media) is the thickest layer, consisting of smooth muscle, collagen, and elastic tissue. This layer is responsible for vascular tone. The outermost layer, the adventitia, consists of connective tissue that provides passive structural support. Blood supply to the wall itself comes from vasa-vasorum, which are small arteries that enter through the adventitia, arborize, and terminate near the junction of middle and outer thirds of the media.[2,26] By way of the vasa-vasorum, the coronary and brachiocephalic arteries perfuse the ascending aorta, the intercostal arteries perfuse the descending thoracic aorta, and the lumbar and mesenteric arteries supply the abdominal aorta.[7,27]

By convention, the aorta is divided into segments[2] (Figs 1 and 2). The ascending thoracic aorta extends from the aortic annulus to the brachiocephalic (or innominate) artery. The proximal portion, referred to as the aortic root, spans the annulus to sinotubular junction and includes the aortic valve and coronary artery ostia. The aortic arch gives rise to the head and neck vessels starting at the brachiocephalic artery and ending at the left subclavian artery. The descending aorta extends from the left subclavian artery to the iliac bifurcation and is subdivided into the descending thoracic aorta and the abdominal aorta, which give rise respectively to the intercostal and brachial arteries, and the mesenteric, renal and lumbar arteries.

The normal aortic wall thickness is < 3 mm.[2] The aortic diameter varies between segments and by age, sex, and patient

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Table 1
Acute Aortic Syndromes.

Lesion	Layer affected	Demographics	Presentation	Location	Appearance	Complication
IMH	Medial layer	65-70 y HTN Males Iatrogenic Trauma	AAS	Descending > Ascending > Arch	Crescentic thick Circumferential thick Varying length No intimal flap No false lumen	Rupture Dissection Aneurysm
AD	Intimal tear Intimal flap Medial false lumen	58-63 y HTN CTD Coarctation Bicuspid AoV Pregnancy Trauma • Iatrogenic	AAS	Ascending > Descending	Intimal flap Dual lumen: True/False Low-flow false lumen False > True lumen	Peripheral ischemia Embolization Aneurysm Rupture
AA	Intima Media Adventitia	50-60 y Males	Asymptomatic Compression of proximal structures Fistula related	Ascending > Descending > > > Arch	Dilation No intimal flap No dual lumen No false lumen	Rupture Embolization Fistula
PAU	Intimal layer	70 y Males HTN Tobacco CAD COPD	AAS	Descending > > > Arch > > Ascending	Irregular surface Crater-like protrusion No intimal flap No dual lumen No false lumen	IMH Aneurysm Embolization Dissection Rupture
ULP	Intimal layer	65 y Males	Asymptomatic	Distal Arch Proximal descending > > > Ascending and Distal Descending	Single Contrast-Filled outpouchings across intima into medial layer No connections with aortic branches	Aneurysm Regression
IBP	Medial layer	60-62 y Males	Asymptomatic	Descending > > > Arch > > Ascending	Multiple pools medial layer No communication Near branch vessels	Disappear

NOTE. Adapted from various references.[1–25]
Abbreviations: AA, aortic aneurysm; AAS, acute aortic syndrome; AD, aortic dissection; AoV, aortic valve; CAD, coronary artery disease; COPD, chronic obstructive pulmonary disease; CTD, connective tissue disorder; HTN, hypertension; IBP, intramural blood pool; IMH, intramural hematoma; PAU, penetrating atherosclerotic (aortic) ulcer; ULP, ulcer-like projection.

size. Aortic diameters are larger in older men with higher body surface areas[2,28] (Table 2). Except in very large individuals, a normal the aortic diameter is rarely >4 cm. An ascending aorta >4 cm is abnormal, and >5 cm is aneurysmal, whereas a descending aorta >4 cm is considered aneurysmal.[2,26] Normalized mean values and upper limits of normal for adults are 1.8 ± 0.2 cm/m^2 and 2.1 cm/m^2 in the ascending aorta, and 1.4 ± 0.2 cm/m^2 and 1.8 cm/m^2 in the descending aorta[2,28] (Fig 1).

Intramural Hematoma

Aortic IMH was first described by Kruckenberg in 1920 as "dissection without intimal tear" that results from hemorrhage within the aortic wall[31] (Fig 3). It has long been suggested that an IMH results from the spontaneous rupture of vasa-vasorum in the medial layer due to a combination of wall stress, fragile vessels, chronic hypertension, and inflammation.[5,14–16] The lack of a clear communication between the lumen and the aortic wall at imaging or surgery is the hallmark of spontaneous IMH. Hemorrhage within the wall causes smooth, crescentic or circumferential mural thickening ($\geq$5 mm) at cross-sectional imaging (Figs 4 and 5). The longitudinal extent of the IMH can be very short ($\sim$1 cm) or can extend the full length of the aorta. In the case of type A IMH, aortic wall thickening creates a separation between the aortic lumen and the right atrial appendage where such a separation normally does not exist (Figs 4–6). This anatomic occurrence helps to distinguish IMH from other AAS.

Rarely with IMH does mural thickening sufficiently narrow the vascular lumen to impair blood flow or compromise branch vessel flow.

More recently, perhaps due to improved imaging technology and resolution, small intimal tears have been detected in as many as 70% to 80% of IMH cases, and may represent a site of initial insult.[32–36] In a report of IMH, intimal tears were