Aortitis Masquerading as Intramural Hematoma: When to Observe, When to Operate? A Case Report

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Abstract

Radiologic evidence of aortic disease is not always consistent with the diagnosis. With lack of accompanying symptoms or with an atypical presentation, diagnosis of aortic pathology relies greatly on imaging techniques. We report the case of a 58 year-old female who presented with incidental radiographic findings consistent with a type A aortic intramural hematoma and a vague left-sided chest discomfort. After follow-up imaging was consistent with disease progression and hematoma expansion, the affected segment was resected and pathology reported lymphoplasmacytic aortitis as the underlying etiology of the imaging findings rather than an intramural hematoma. The patient lacked symptoms or serology consistent with rheumatologic disease and the postoperative course was uneventful. The management of a suspected ascending intramural hematoma is controversial, especially when the patient presents with atypical signs and symptoms. Features of disease progression may warrant urgent surgical intervention.

Case report

Aortic intramural hematoma (IMH) represents 6.3% of all acute aortic syndromes and most commonly involve the descending aorta.[1] It occurs predominantly in elderly males with hypertension and is presumed to be caused by rupture of the vasa vasorum, pathological neovascularization, or bleeding originated by rupture of an ulcerated plaque.[2] It usually presents with abrupt chest or back pain and diagnosis is based on computed tomography or transesophageal echocardiography.[2]

In contrast to the management of aortic dissection (AD), the standard treatment for IMH and clinical course remains unclear.[3] As with AD, IMH is often categorized as either Stanford type A or B. Conservative management is often the strategy for type B IMH. However, much controversy exists for the treatment of type A IMH, especially in patients with aspecific symptoms such as with the case presented. Follow-up imaging studies and expedite surgical repair in those who demonstrate hematoma expansion has been suggested as the standard of care.[4]

A 58 year-old female with history of hypertension, dyslipidemia and diabetes mellitus was referred to our emergency department following a CT scan of the neck obtained for the workup of a parathyroid adenoma. During review of the images an aortic intramural abnormal finding, measuring 0.9 cm in thickness and with 73 Hounsfield density units (consistent with the density of a hematoma: 40-90 HU), was incidentally surrounding the ascending thoracic aorta. A small pericardial effusion with identical radiologic density was associated with the findings. The patient denied chest or back pain except for occasional self-resolving discomfort in the left infrascapular region.

Six month follow-up chest CT angiography was consistent with an increase in size of the hematoma (2.7 cm), now involving the ascending aorta and the proximal aortic arch with a new focal protrusion at the level of the main pulmonary artery with accompanying small-to-moderate hemopericardium (**Fig. 1A and B**). There was no evidence of dissection or aneurysm, no concomitant severe coronary artery calcification

and the rest of the aorta and its branches were normal. A transthoracic echocardiogram showed no valve abnormalities and a well-preserved left ventricular function with an ejection fraction estimated at 60%. In light of the progression of the hematoma and extension into the main pulmonary artery, urgent surgical intervention was recommended with plans for replacement of the involved ascending aorta.

Upon entry into the pericardium, the expected hemopericardium turned out to be a clear effusion. The ascending aorta had a leather-like aspect with white dense fibrotic tissues surrounding the adventitia and a thickened aortic wall extending into the arch (**Fig. 2A and B**). There were dense fibrous inflammatory adhesions between the medial aspect of the ascending aorta and the pulmonary artery. Under cardiopulmonary bypass and deep hypothermic circulatory arrest, the distal ascending aorta was resected. The wall appeared to be severly thickened with dense fibrotic tissue, and there was no evidence of intimal entry tears, flaps or an intramural hematoma. The diseased aortic segment was replaced with a 26 mm Dacron graft.

Pathology of the specimen revealed extensive lymphoplasmacytic infiltrate in an expanded, markedly fibrotic adventitia consistent with lymphoplasmacytic aortitis (**Fig. 3A and B)** . The lymphoid infiltrate was found to be polymorphous, with a mild predominance of B-cells (**Fig. 3C**). Staining for Ig showed predominance of IgG positive cells (**Fig. 3D**) but only a few IgG4, ruling out IgG4 related disease. Further serologic assessment found no evidence of auto-antibody-associated disease markers and treponemal serologies were negative as well.

The management of intramural hematoma remains unclear, especially for those who have an asymptomatic presentation or an incidental finding. Hata M. et al[3] reported a postoperative mortality of 5.4% for those with type A IMH complicated by cardiac tamponade, aortic regurgitation, acute MI or organ malperfusion undergoing emergency surgery, 0% for those with uncomplicated IMH who had emergency surgery and 25.8% for those had conservative management. This significant difference in mortality supports the aggressive treatment for type A IMH with a rationale that uncomplicated patients are hemodynamically stable prior to surgery, decreasing perioperative mortality significantly.[3]

Unfortunately, as this case shows, oftentimes etiology cannot be determined solely on imaging techniques despite the predictive accuracy that radiologic features may have, and intra-operative findings may not be consistent with disease presentation. When there is suspicion for aortitis, positron emission tomography (PET) could potentially aid in differentiating it from IMH.[5] However, this patient lacked symptoms of rheumatologic disease, inflammatory markers or other features suggestive of lymphoplasmacytic aortitis that would warrant such imaging at the time.

The presented case demonstrates the discrepancy between radiologic features (expanding type A IMH) supporting surgical intervention, and intraoperative and pathology findings that could have supported a conservative management. It remains largely up to the surgeon's discretion to make a decision to operate in early stages or to monitor progression to features that may warrant urgent care such as expanding aneurysm, involvement of other structures, hemopericardium or risk of dissection.

References:

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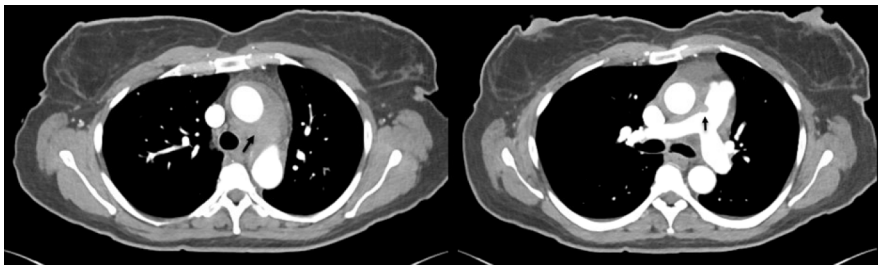
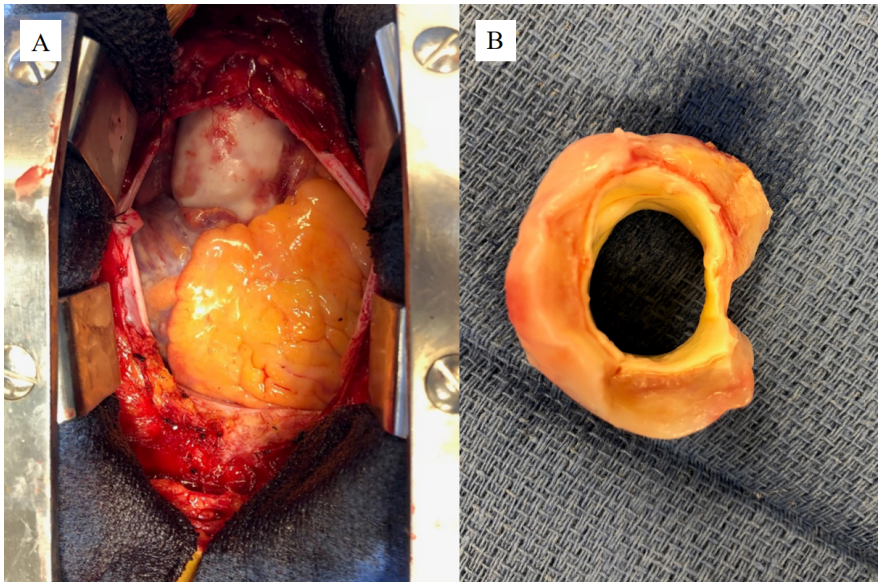
Figure legends

Figure 1: *Preoperative chest CT scan*



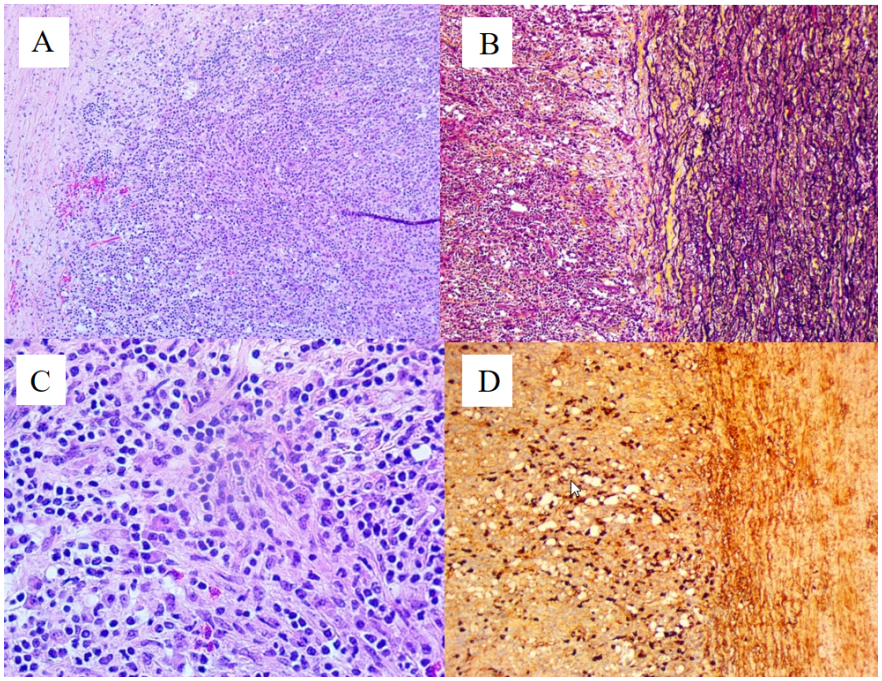
Legend: Intramural hematoma in the aortic arch (A) with enlargment and protrusion into the pulmonary artery, before surgery (B).

Figure 2: *Operative view and gross findings of the ascending aorta*



Legend: Intraoperative findings demonstrating the ascending aorta with a leatherlike appearance (A). Cross section of diseased segment with circumferential thickening of the aortic wall (B).

Figure 3: *Histopathology of the aortic specimen consistent with lymphoplasmacytic aortitis*



Legend: Pathology report of the excised segment demonstrating inflammatory infiltrate in the aortic wall (4x) (A) affecting predominantly the adventitia (10x) (B). Polymorphous inflammation (20x) (C) with IgG positive plasma cells (10x) (D).