

## IgG4-related aortitis mimicking intramural hematoma

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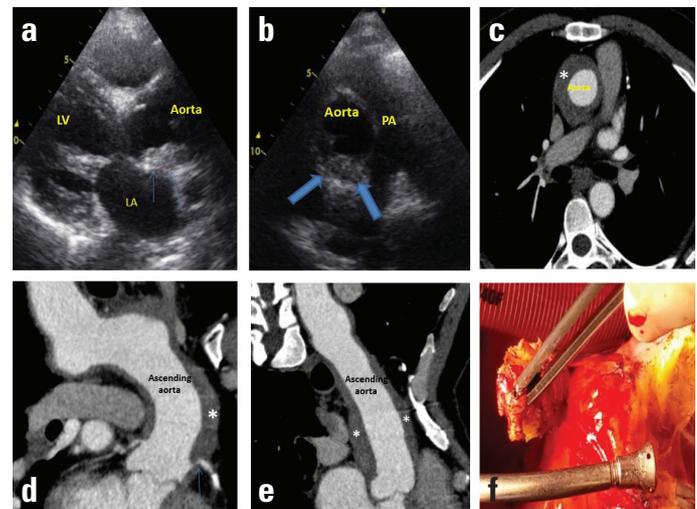
### Introduction

Acute aortic syndromes (AAS) are life-threatening conditions, and despite advances in imaging techniques, their diagnosis and treatment remain challenging. While intramural hematoma is much less common than acute aortic dissection (AD), which is the most common form of AAS, they have similar mortality rates (1). On the other hand, aortitis may mimic intramural hematoma (IMH) due to radiologic similarity and, thus, may lead to misdiagnosis and treatment.

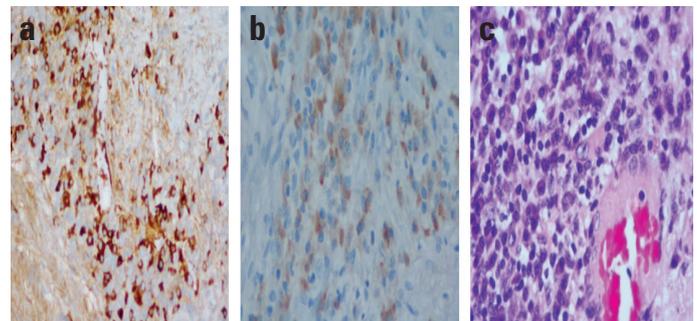
Here, we describe an IgG4-related aortitis case mimicking intramural hematoma (IMH) due to radiologic image and presenting with chest pain and severe aortic regurgitation.

### Case Report

A 49-year-old male patient with new-onset retrosternal chest pain was admitted to our clinic, and unstable angina was suspected. He had a history of coronary artery by-pass grafting surgery 7 years ago and stenting for right coronary artery 1 year ago. He was under statin treatment for hyperlipidemia. On physical examination, his heart rate was 110/min, blood pressure was 104/45 mm Hg, and an aortic diastolic murmur was auscultated. Electrocardiography was free from signs of new ischemia. Echocardiographic examination revealed normal left ventricle systolic function, ascending aortic aneurysm with maximal diameter of 4.7 cm, and severe aortic regurgitation, which were not present in previous exams. Aortic wall thickness was visually noted to be increased in parasternal long-axis (Fig. 1a). A preliminary diagnosis of AAS was established using computed tomography (CT). CT images were consistent with a circular IMH image with a thickness of 12 mm and extending from aortic root to aortic arch and also containing orifices of major branches of aorta (Fig. 1b, c). No entry for flap or dissection was seen. CT coronary angiography showed normal circumflex artery, patent LIMA-AD, and IMH containing ostia of both left main coronary artery and right coronary artery RCA. Moreover, external pressure by IMH caused LMCA stenosis >50%. Heart team recommended surgery to the patient with ongoing chest pain. Re-sternotomy was performed through an old incision line. Because of unexpectedly severe adhesive and firm character of tissues in the surgical area, the ascending aorta, branches of aortic arch, and thoracic aorta could not be clearly distinguished during surgery. Aortic lumen was reached



**Figure 1.** (a,b) Parasternal long and short-axis images showing thickened aortic wall (blue arrows) starting just above aortic valve (c,d,e) CT image showing circular thickened aortic Wall (\*) extending from aortic valve level to aortic arch and mimicking intramural hematoma image, and the relationship of coronary ostium with wall thickness (blue arrow), (f) macroscopic thickened aortic wall image obtained from intraoperative specimens



**Figure 2.** Lymphoplasmocytic infiltration in biopsy material obtained from aorta (a) Immunohistochemical IgG staining (original magnification x20), (b) Stain acquisition with IgG4 in 50% of the cells stained with immunohistochemical IgG (x20), (c) Hematoxylin and eosin staining showing inflammation including lymphocytes and plasma cells but no granuloma formation (lymphoplasmocytic pattern) (x40)

by cutting the stiff tissue around the aortic outer surface, and aortic wall thickness was measured as 11 mm. A 23-mm St. Jude's prosthetic valve was implanted with Ti-Cron suture. After 2 days, during follow-up in the coronary care unit, the patient died. Pathological examination of the aorta revealed IgG4-related aortitis and chronic inflammation containing medial, adventitial, and periadventitial lymphocytes and plasma cells (Fig. 2a-c).

### Discussion

IgG4-related aortitis is defined as an inflammatory and non-infectious type of vasculitis. The exact mechanism underlying this pathology remains to be elucidated; however, genetic profile and autoimmune mechanism triggered by bacterial infections

have been implicated. Estimated prevalence of IgG4-related thoracic aortitis is 9–22% (2). It is associated with increased erythrocyte sedimentation rate, C-reactive protein, and Ig-G4 levels in plasma (2).

Long-term mortality of IMH is similar to that of aortic dissection; however, it differs from dissection with bleeding into the aortic wall and absence of false lumen and intimal tear. It constitutes 10–25% of AAS cases. IgG4-related aortitis is a relatively uncommon form of aortitis, which may mimic IMH and usually affects individuals older than 50 years. Moreover, it can cause pancreatitis, sialadenitis, lymphadenopathy, sclerosing cholangitis, nephritis, and retroperitoneal fibrosis via systemic inflammatory response. According to 2015 dated consensus statement of Society for Cardiovascular Pathology, the requirements for the diagnosis of IgG4-related aortitis/periaortitis were increased levels of serum IgG4, histopathologically confirmed fibrosis, an IgG4/IgG ratio greater than 50%, and more than 50 IgG4+ plasma cells per 400x HPF (3).

IgG4-related aortitis can be confused with IMH because of increased aortic wall thickness, and in this context, various cases have been reported. Tay et al. (4) made a post-operative diagnosis of IG4-related aortitis in a patient with hoarseness who was previously diagnosed as IMH on CT and had undergone surgery for IMH. Moreover, regression in simultaneously increased abdominal aortic wall thickness was noted after prednisolone treatment for 1 month. Rituximab was shown to be effective in regression of refractory or recurrent disease despite steroid therapy. Holmes et al. (5) reported an IgG4-related aortitis case presenting with sudden cardiac death associated with aortic dissection extending into LMCA. Similarly, our patient suffered new-onset chest pain and severe aortic regurgitation due to complications of progressive IgG4-related aortitis. The reasons responsible for greater difficulty during surgery than anticipated were; a) involvement of a wide segment of ascending aorta, b) involvement of coronary arteries thus leading severe aortic regurgitation, c) re-sternotomy, d) lack of being on medical therapy for aortitis. Although early surgical intervention is recommended for Type A IMH, investigation for aortitis mimicking IMH will

change the course of management. Unless patients diagnosed with IMH is unstable, examination of levels of plasma IgG and IgG4 should help establishment of accurate diagnosis.

## Conclusion

Our ability to detect intramural hematoma has increased with the advancement of diagnostic technology and better classification of acute aortic syndromes. Not only morphological but also pathological properties such as inflammatory reaction that develop during these syndromes are very similar to each other. Keeping this fact in mind is critical when deciding on diagnostic and therapeutic strategies.

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