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Plasmacytic Aortitis with Occlusion of the Right Coronary Artery

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Male, 55
Final Diagnosis: Plasmacytic aortitis
Symptoms: Dizziness • nausea
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Unusual clinical course

Background: Inflammation of the aortic wall, known as aortitis, is a rare clinical entity which is frequently asymptomatic, or identified when the patient presents with an aortic aneurysm or dissection. It is most often caused by infection or autoimmune vasculitides such as giant cell or Takayasu's arteritis.

Case Report: The case presented is that of a 55-year-old man with symptomatic occlusion of the right coronary artery caused by a plasmacytic aortitis suggestive of IgG4 disease, which was successfully treated with coronary artery bypass grafting and an ascending aortic graft.

Conclusions: A review of the current literature emphasizes how poorly the etiology and natural history of plasmacytic aortitis is understood.

MeSH Keywords: Aortic Aneurysm • Aortitis • Immunoglobulin G

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Background

Aortitis is a condition characterized by inflammatory infiltration of the aortic wall in response to autoimmune cell signaling [1–4], as seen in Takayasu's arteritis and giant cell arteritis, or with infections such as syphilis [5], salmonella, and tuberculosis. Typically, patients with aortitis have either aortic dissections or aneurysms due to chronic inflammation. A small subset of patients have predominantly plasmacytic aortitis of uncertain etiology. Recently IgG4 related disease has been associated with plasmacytic aneurysmal disease of the ascending aorta [6,7] as well as inflammatory infrarenal abdominal aortic aneurysms (AAA) [8]. This case report describes an atypical presentation of plasmacytic aortitis in which the symptoms were due to inflammatory occlusion of the ostium of the right coronary artery.

Case Report

The patient was a 55-year-old African American man who presented to the Emergency Department of a peripheral hospital with complaints of vague chest pain, nausea, and dizziness. In the Emergency Department he suffered a cardiac arrest secondary to ventricular fibrillation. He was quickly resuscitated and did not have any neurological deficit. CT angiography was suggestive of intramural hematoma of the ascending aorta and arch (Figure 1). He was transferred to our institution for further evaluation.

Cardiac catheterization revealed a severe stenosis of the ostium of the right coronary artery, with otherwise normal coronary arteries (Figure 2). Because the coronary stenosis appeared to be due to compression from the suspected intramural hematoma, he was taken to the operating room for surgical repair. Intra-operatively, the ascending aorta was noted to be firm with a thick wall (Figure 3). There was no evidence of dissection or intramural hematoma. Transection of the ascending

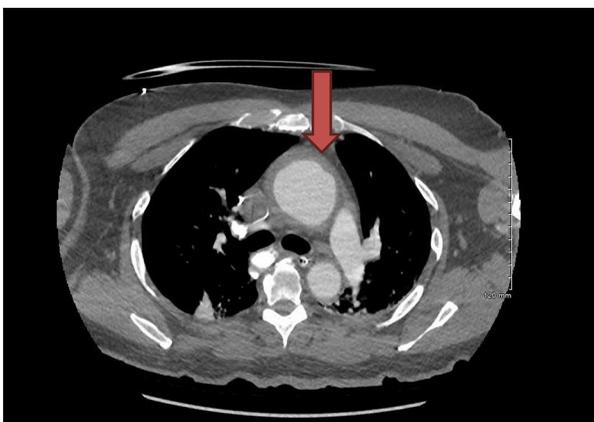


Figure 1. CTA, suggestive of intramural hematoma.

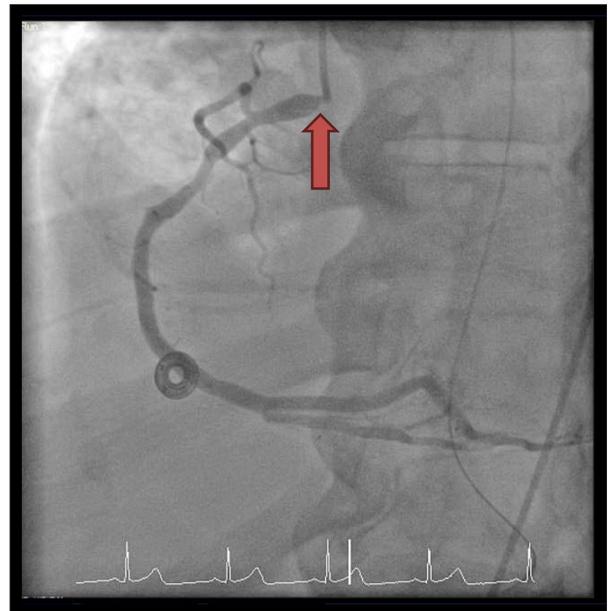


Figure 2. Cardiac catheterization, right coronary artery stenosis.

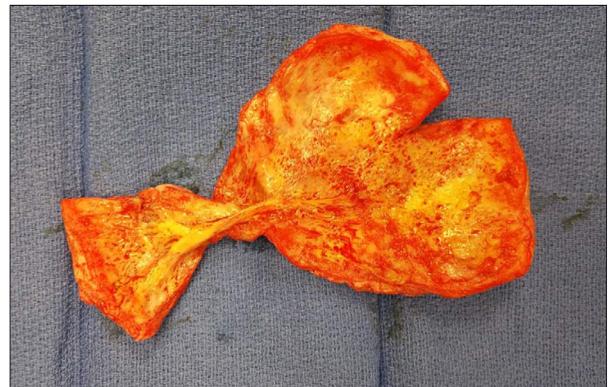


Figure 3. Operative picture, thickened ascending aortic wall.

aorta revealed extreme thickening and edema of the aortic wall consistent with vasculitis. The right sinus of Valsalva also had evidence of inflammation causing severe stenosis at the origin of the coronary artery. The aortic valve and remaining sinuses of Valsalva appeared normal. The ascending aorta was resected and replaced from the sinotubular junction to the innominate artery with a 28-mm Dacron tube graft with end-to-end anastomoses. A saphenous vein bypass graft to the right coronary was then performed. In retrospect, if the diagnosis of aortitis had been clear, perhaps the right coronary stenosis could have been corrected with a percutaneous stent. The patient tolerated the procedure without complications. He recovered rapidly and was discharged on the 7th postoperative day on oral steroid therapy. He returned with a superficial wound infection at the saphenous vein harvest site, which healed well with conservative management. He remains asymptomatic with no evidence of aortic aneurysmal disease by CT scan. Pathology examination of the ascending aorta revealed

aortitis with marked fibrous thickening of the aortic wall and prominent hyaline thickening of vaso vasorum vessels, many of which were surrounded by significant, predominantly plasmacytic, chronic infiltration. Gram stain, fungal cultures, and AFB cultures were all negative. Qualitative and quantitative RPR (rapid plasma reagin) and syphilis IgG and IgM antibodies were also negative. Steiner staining of the aortic wall showed no spirochetes. Tissue staining for IgG4 revealed less than 30% of plasma cells were positive for IgG4. Serum IgG4 levels were normal, but the laboratory specimen was collected after treatment with oral steroids.

Discussion

Aortic aneurysms involving all segments of the aorta from the ascending to the infrarenal anatomy have long been recognized and attributed to a variety of autoimmune etiologies including Takayasu's arteritis and giant cell arteritis and other rheumatologic diseases such as rheumatoid arthritis, Behçet's disease, and Cogan's syndrome [1–5]. However, a significant minority of cases (<10%), usually classified as idiopathic, are associated with predominantly plasmacytic infiltrates rather than macrophages and lymphocytes. Some of these cases may be related to IgG4 disease but the etiology of other cases remains obscure [6–8].

IgG4-related aneurysmal disease has recently been identified in aneurysms affecting all segments of the aorta and may be an important etiology of inflammatory infrarenal AAA. IgG4-related disease has been identified with aneurysms of the thoracic aorta, but the criteria to establish the disease remain vague and undetermined [9,10]. All cases demonstrate

plasmacytic infiltrates with greater than 30% of plasma cells staining positively for IgG4, but not all patients have elevated serum levels of IgG4 [11]. To further confuse the diagnosis, IgG4-positive plasma cells have been identified in some rare cases of infective aortitis [12,13].

Patients with IgG4-related thoracic aortic aneurysm disease have not been well characterized but appear to be older compared to the typical patient with vasculitis (>55 years), with no sex predilection and no other features of IgG4 systemic disease. When surgical intervention is not urgent, there is some evidence that steroid therapy may result in regression of disease [14–16]. The etiology of plasmacytic aortitis without evidence of IgG4 staining or elevated plasma levels is unclear. Plasmacytic infiltrates may represent a response to a variety of inflammatory etiologies, or the current methods of identifying IgG4 disease may be inadequate.

This reported case is an unusual presentation of idiopathic plasmacytic aortitis of the ascending aorta, possibly related to IgG4 disease, who presented with sudden death/ventricular fibrillation secondary to right coronary artery stenosis. This patient, however, did not have elevated IgG4 markers. Five similar cases that culminated in death have been reported in the literature [17–21].

Conclusions

The understanding of the etiology, diagnosis, natural history, and response to therapy for plasmacytic aortitis aneurysmal disease are poor. Case report studies and further evaluation are necessary to advance our understanding of this pathology.

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