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Case Report

The great imitator: IgG4 periaortitis masquerading as an acute aortic syndrome on computed tomographic angiography

Drew W. Moore DO^a, Neil J. Hansen MD^{a,*}, Dominick J. DiMaio MD^b,
William L. Harrison MD^a

^a Department of Radiology, University of Nebraska Medical Center, 981045 Nebraska Medical Center, Omaha, NE 68198-1045, USA

^b Department of Pathology, University of Nebraska Medical Center, Omaha, NE, USA

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ABSTRACT

We present the case of a 52-year-old woman who presented to the emergency department with chest and neck pain. Initial cervical spine magnetic resonance imaging shows an abnormal flow void in the left vertebral artery, which prompted a computed tomographic angiogram. This demonstrated a hyperdense thickened ascending aortic wall, which extended into the great vessel origins. Clinically and radiographically interpreted as an acute aortic syndrome and/or intramural hematoma, the patient underwent ascending aortic repair with graft. An unusual aortic and/or periaortic mass was encountered in surgery and final pathology demonstrated IgG4 periaortitis. A rare clinical disease, IgG4-mediated processes are often mimickers of other pathologic entities and frequently lead to misdiagnosis. All pathologically similar, IgG4-mediated disease processes can involve the pancreas, salivary glands, orbits, retroperitoneum, and the vasculature.

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Case report

A 52-year-old woman with a medical history of gastroesophageal reflux, hiatal hernia, and hypertension presented to the emergency department with ongoing intermittent chest and neck pain. She stated that 10 days before admission, she experienced severe chest and left shoulder pain with associated temporary loss of left arm function. In addition, her entire arm temporarily turned gray and dusky. While these

arm symptoms resolved, the neck and chest pain continued and prompted her to seek medical care. On initial presentation to the emergency room, her physical examination was entirely normal.

Due to concern that her clinical symptoms were related to a compressive neuropathy, cervical spine magnetic resonance imaging was obtained (Fig. 1). This magnetic resonance imaging demonstrated an abnormal lack of flow void in the left vertebral artery. Subsequently, computed tomographic

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* Corresponding author.

E-mail address: njhansen@unmc.edu (N.J. Hansen).

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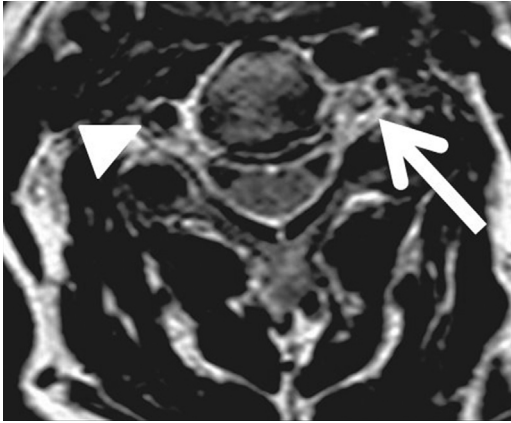


Fig. 1 – Axial T2-weighted image through the cervical spine demonstrates an abnormal lack of flow void in the left vertebral artery, which is high signal (arrow). The right vertebral artery (arrowhead) demonstrates a normal flow void.

(CT) angiography of the head, neck, and chest (Fig. 2) revealed hyperdensity and thickening of the ascending aorta and proximal arch aortic wall. Extension into the origin of the great vessels resulted in near complete occlusion of the left vertebral artery (Fig. 3). The remainder of the arterial vasculature, including the descending thoracic aorta, the abdominal aorta, and all major branch vessels were widely patent and normal with no wall thickening. All abdominal parenchymal organs were normal. Specifically, the pancreas demonstrated normal morphology with no enlargement or other features of autoimmune pancreatitis.

On imaging, the differential diagnosis for aortic wall thickening is limited. Given the mild hyperdense appearance on the noncontrast portion of the CT obtained, and the clinical suspicion for an acute aortic pathology, the leading differential diagnosis was an acute intramural hematoma. Occasionally, an aortic dissection with a thrombosed false lumen can have a similar imaging appearance (but it is managed similarly so imaging distinction is unimportant). Infectious and inflammatory vasculitides can cause aortic wall thickening and appear similar on imaging. Clinically, these entities



Fig. 2 – Axial (A) and coronal oblique (B) noncontrast CT images demonstrate thickening and subtle hyperdensity of the aortic wall involving the ascending aorta and proximal arch (white arrow and arrowhead). Postcontrast CT angiogram confirms the presence of aortic wall thickening (up to 7 mm—black arrowhead in C), which extended up along the walls of the brachiocephalic (black arrow—D) and left subclavian arteries.

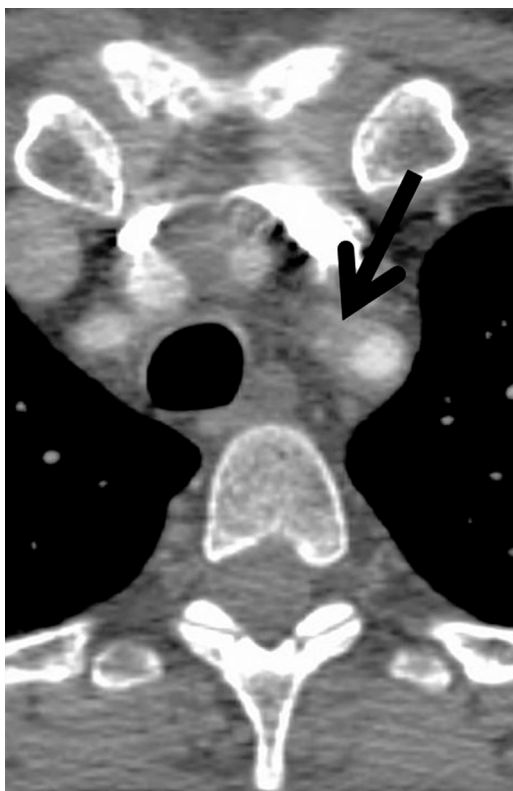


Fig. 3 – Axial contrast-enhanced CT angiogram image at the level just above the great vessel origins demonstrates near occlusion of the left vertebral artery (arrow) just after its takeoff related to aortic wall thickening.

usually have a more insidious onset of symptoms without an acute component as was seen in this case. Aortic wall neoplasms (typically sarcomas) are exceedingly rare and usually have more of an irregular intraluminal or exophytic mass-like morphology.

Given the clinical presentation and imaging findings, the patient went to the operating room for repair of a presumed acute aortic syndrome involving the ascending aorta. Transesophageal echocardiography done during the median sternotomy identified a possible intraluminal flap in the ascending aorta. During the operation, concentric blue mass-like hard thickening was observed to involve the distal ascending aorta and proximal arch. Surgical repair of the ascending aorta was done with placement of a 26-mm tube graft. The patient tolerated the procedure well, and postoperative CT angiography demonstrated no postoperative complications (Fig. 4). Pathologic evaluation demonstrated a lymphoplasmacytic inflammatory infiltrate involving the entire thickness of the ascending aortic wall (Fig. 5). Fibrosis was present in the adjacent periaortic soft tissues. A predominance of B-cells was found on immunophenotyping with an increased ratio of IgG4 in relation to other immunoglobulins. The final diagnosis was IgG4-related periaortitis. Her serum IgG4 level was normal at 50 mg/dL (normal serum levels are seen in up to 15% of patients with IgG4-related diseases). The patient was treated with systemic immunosuppressive drugs including prednisone, rituximab, and

methotrexate, but during the course of her therapy discontinued immunosuppression for perceived drug side effects and is currently not on therapy and asymptomatic. Three postoperative CT scans (1, 12, and 18 months postoperative) have remained stable while off therapy with no new changes.

Discussion

IgG4-related diseases (IgG4-RD) were first recognized in autoimmune pancreatitis and have been identified in numerous other organ systems [1–3]. Organs that can be involved include the salivary glands, thyroid, mediastinum, heart, kidneys, retroperitoneum, and the large vessels such as the aorta. IgG4-related periaortitis is a pathologic diagnosis that has only been described in the last decade, predominately in case reports and small series of cases. The pathology is typically described as being similar to other IgG4-RD, with lymphoplasmacytic infiltrate enriched fibrosis being the dominant feature. This has been termed storiform fibrosis due to its morphology. Periaortitis has been the preferred term over aortitis, and the following criteria are used: histologic appearance consistent with aortitis or periaortitis and not explained by other disease processes (eg, atherosclerosis), at least 50% plasma cells stain for IgG4, and at least 50% of cells are IgG4 plasma cells per high-powered field [3]. Given IgG4 plasma cells are elevated, serum IgG4 levels are typically but not always increased. This is not a specific finding; however, as serum IgG4 levels can be increased in a variety of infections and neoplastic processes.

There are only a few other reports similar to this, where IgG4 periaortitis was mistaken for an acute aortic syndrome, and a patient went to the operating room [4–6]. In these cases, similar operative findings were encountered in terms of the aorta and adjacent tissues having a blue or whitish tinge and being firm to palpation. Conventionally, autoimmune-mediated vasculitides are treated with immune modulation and suppression rather than operative intervention. However, diagnostic and clinical management dilemmas occur when acute presentations involve anatomic locations that are usually treated with surgery (eg, ascending aorta). To complicate matters further, the inflamed vascular wall may have a propensity toward concomitant acute aortic pathology, as an acute dissection related to IgG4 periaortitis has been reported [7].

There is no standard treatment for IgG4 periaortitis as of yet. In some reports, when operative findings were not typical for an intramural hematoma, surgical repair was aborted, and the patient was successfully treated with steroids [6]. Current case reports and/or series and clinical trials are evaluating the efficacy of traditional immunomodulatory drugs such as steroids and methotrexate, although newer agents such as the anti-CD20 antibody rituximab are also being studied to treat these autoimmune vasculitides [5,8,9].

Precise imaging signs that may differentiate an acute intramural hematoma from an inflammatory periaortitis have yet to be fully elucidated—and in cases this differentiation may be impossible with imaging. Given the risks of not treating an intramural hematoma involving the ascending aorta, surgical intervention in cases such as this may occur



Fig. 4 – Postoperative axial contrast-enhanced CT angiography images (A and B) demonstrate an uncomplicated appearance of the ascending thoracic aorta status after repair. Arrows demonstrate surgical material at the origin and touchdown sites of the surgical graft. Coronal oblique precontrast (C) and postcontrast (D) images show the length of the graft (arrowheads), which extended for the entire length of the ascending aorta to the proximal arch.

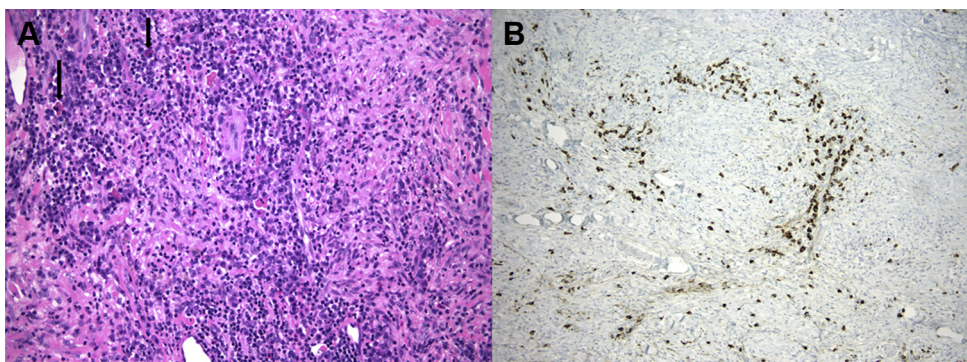


Fig. 5 – (A) Within the aorta and adjacent soft tissue, there is a lymphoplasmacytic inflammatory infiltrate. A prominent number of plasma cells and Mott cells (black arrows) are present (H&E, 200 \times). Mott cells are plasma cells that have multiple cytoplasmic inclusions (called Russell bodies) that represent immunoglobulins. (B) Immunohistochemistry staining for IgG4 demonstrated increased expression within the plasma cell population (IgG4, 100 \times).

when clinical symptoms are acute. It is up to the radiologist to raise the possibility of vasculitis and periaortitis in these scenarios. Further investigation into this area is needed to investigate whether imaging may be able to differentiate these entities and avoid unnecessary surgery.

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