Radiology Case Reports xxx (2016) 1–5

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Case Report

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

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ARTICLE INFO

Article history:
Received 17 May 2016
Received in revised form
12 July 2016
Accepted 12 August 2016
Available online xxx

Keywords:
Acute aortic syndrome
IgG4
Aortitis
Intramural hematoma
CT angiography
Periaortitis

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.

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

Competing Interests: The authors have declared that no competing interests exist.
Consent: The consent was obtained directly from the patient involved before submission of this manuscript.
Human and animal rights: Not applicable to this case report.

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http://dx.doi.org/10.1016/j.radcr.2016.08.006

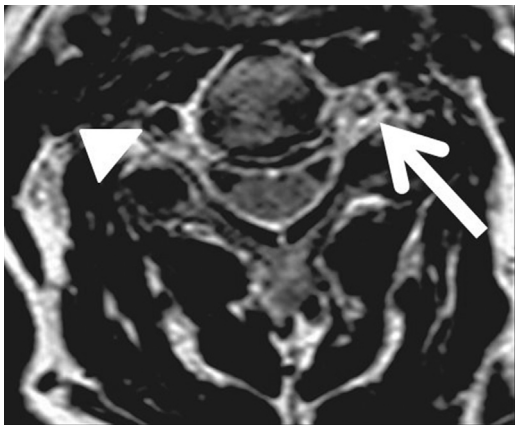

Fig. 1 – Axial T2-weighted image through the cervical spine demonstrates an abnormal lack of flow void in the left vertebral artery, which is high signal (arrow). The right vertebral artery (arrowhead) demonstrates a normal flow void.

(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

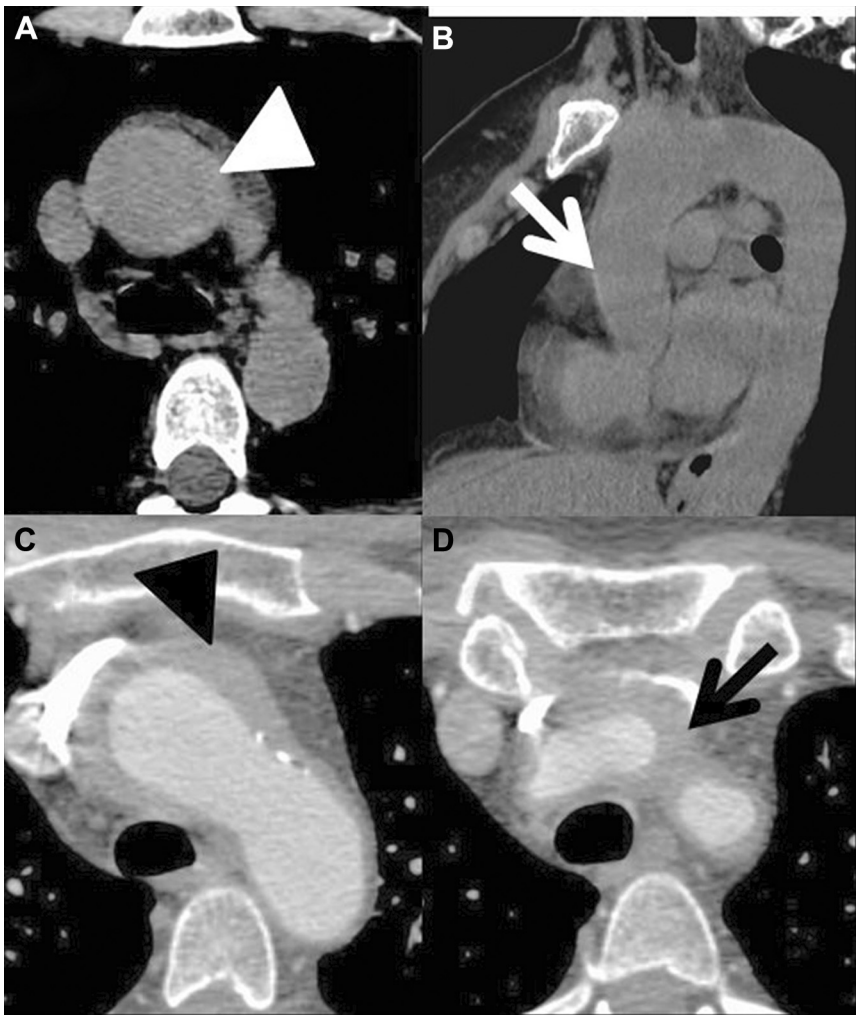

Fig. 2 – Axial (A) and coronal oblique (B) noncontrast CT images demonstrate thickening and subtle hyperdensity of the aortic wall involving the ascending aorta and proximal arch (white arrow and arrowhead). Postcontrast CT angiogram confirms the presence of aortic wall thickening (up to 7 mm—black arrowhead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