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## International Journal of Surgery Case Reports

journal homepage: [www.casereports.com](http://www.casereports.com)

## Using temporal artery biopsy to diagnose giant cell arteritis in a patient with bilateral arm ischemia

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## ARTICLE INFO

## Article history:

Received 31 March 2015

Accepted 9 June 2015

Available online 19 June 2015

## Keywords:

Giant cell arteritis

Upper extremity ischemia

Claudication

## ABSTRACT

**INTRODUCTION:** Bilateral upper extremity ischemia is an unusual presentation of vascular disease. Aetiologies include atherosclerosis as well as rheumatologic diseases. History and physical examination are often, but not always, enough to distinguish between aetiologies and guide treatment.

**PRESENTATION OF CASE:** We present the case of a female patient with findings neither typical for atherosclerotic or for rheumatologic disease who was ultimately found to have giant cell arteritis affecting her bilateral upper extremities. She underwent bilateral upper extremity bypasses using saphenous vein grafts.

**DISCUSSION:** This patient presented without symptoms and laboratory findings often seen with GCA, however, biopsy revealed a definitive diagnosis. Treatment options for ischemia secondary to giant cell arteritis are not well-documented in the literature.

**CONCLUSION:** Giant cell arteritis can present in atypical forms, and should remain on the differential when atypical-appearing lesions are found, even in the absence of features usually associated with GCA.

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## 1. Introduction

The differential diagnosis for upper extremity ischemia is broad and includes atherosclerotic disease, anatomic anomalies, radiation-induced intimal hyperplasia, and large vessel vasculitides. We present a patient who was ultimately determined to have giant cell arteritis (GCA) causing bilateral upper extremity ischemia; her atypical presentation illustrates the importance of a broad differential for upper extremity claudication. She consented to the publication of this manuscript.

## 2. Case report

A 69 year old woman presented to the office with bilateral hand pain, numbness, and tingling. She first began to experience exertional arm pain two months prior, and the pain had progressively worsened. She denied any headaches, visual changes, jaw claudication or shoulder or hip girdle stiffness. She had no history of coronary or peripheral vascular disease.

Her only medications were a lisinopril/hydrochlorothiazide combination pill and a statin. She had never smoked and was a vegetarian. Her only past surgery was a bilateral radical mastectomy for breast cancer 25 years prior to presentation; this was accompanied by chemotherapy but not radiation. She had no history of axillary lymph node dissection, radiation or trauma.

On examination, she was a thin, well-appearing woman. She had non-palpable radial and ulnar pulses bilaterally and unobtainable upper extremity blood pressures. She had no temporal artery tenderness or visible pulsation; there was no stiffness with range of motion in her shoulders or hips.

The patient had previously seen her primary care provider and brought a CT angiogram of the neck, which showed a left subclavian artery irregularity with a stenosis proximal to where it crossed the first rib, a focal dilation in the proximal left axillary artery, and severe axillary and brachial artery stenosis. She had an irregularity of her right subclavian artery and an occluded right axillary artery (Fig. 1). She was admitted to the hospital. Labs revealed a mildly elevated ESR (40 mm/h; normal 0–30 mm/h), a normal hemoglobin (12.3 g/dL; normal 12.0–16.0 g/dL) and were otherwise unremarkable.

An angiogram showed a 3–4 cm focal occlusion of her right axillo-brachial artery with reconstitution of her brachial artery (Fig. 2). The left subclavian artery was occluded just past the takeoff

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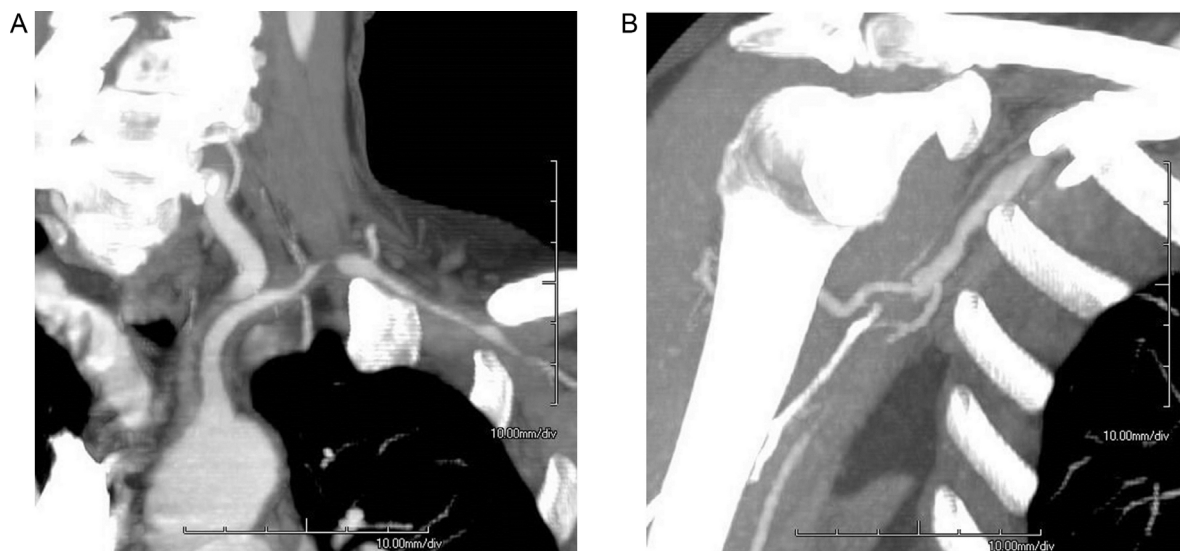


Fig. 1. CT angiogram showing (A) irregularity of the left subclavian and segmental stenoses of the left axillary artery and (B) focal occlusion of the right axillary artery.

of the vertebral. Her left axillary artery reconstituted inferior to the clavicle.

Rheumatology was consulted and recommended biopsy of either the bypass site or temporal artery. A hsCRP was found to be markedly elevated at 118.4 mg/L (normal range <7.4 mg/L). Her lipids were within normal range.

She underwent a left common carotid to left brachial bypass using cephalic vein. A segment of the carotid artery was sent for pathology and showed connective tissue with myxoid changes. She subsequently underwent a right axillary to brachial artery bypass with reversed cephalic vein. The bilateral temporal arteries were biopsied, and pathology showed chronic inflammation with rare giant cells in the media and adventitia, as well as focal disruption of the elastic lamina (Fig. 3). The diagnosis of giant cell arteritis was made, and she was started on prednisone 60 mg daily with a plan to taper over the next eighteen–twenty-four months. Additional imaging of abdominal vasculature revealed critical right renal artery stenosis. Her lisinopril was discontinued. She was discharged home on post-operative day 5 after the second operation. In follow-up in clinic, she felt well with complete resolution of her symptoms.

### 3. Discussion

Giant cell arteritis is found in 7–20 people per 100,000 per year, often in those older than 50 years and of Northern European descent [1]. Its pathogenesis is thought to be a combination of environmental exposures and genetic polymorphisms. CD4 T cells migrate to the vasa vasorum, stimulating inflammation and attracting macrophages, leading to the formation of multinucleated giant cells and intimal expansion [1]. The presentation may be acute or insidious, and includes jaw claudication, as well as constitutional symptoms such as weight loss, fever, and fatigue. Symptoms often include findings of polymyalgia rheumatica, such as shoulder and hip girdle stiffness. Lab findings may include an ESR>80, an elevated CRP, leucocytosis, anemia, and thrombocytosis. The presence of 3 or more American College of Rheumatology Criteria (development of symptoms at the age of 50 or older, headache, temporal artery tenderness or decreased palpation, ESR >50, and biopsy with giant cells) is greater than 90% sensitive and specific [2] for a diagnosis of giant cell arteritis. This patient had none of the typical findings, and did not meet ACR criteria for giant cell arteritis.

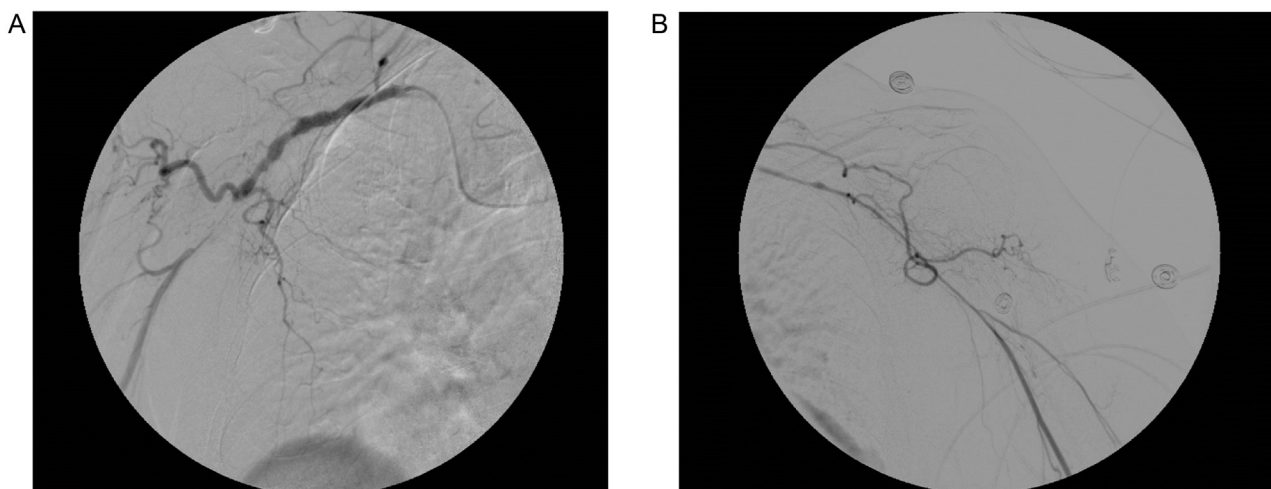
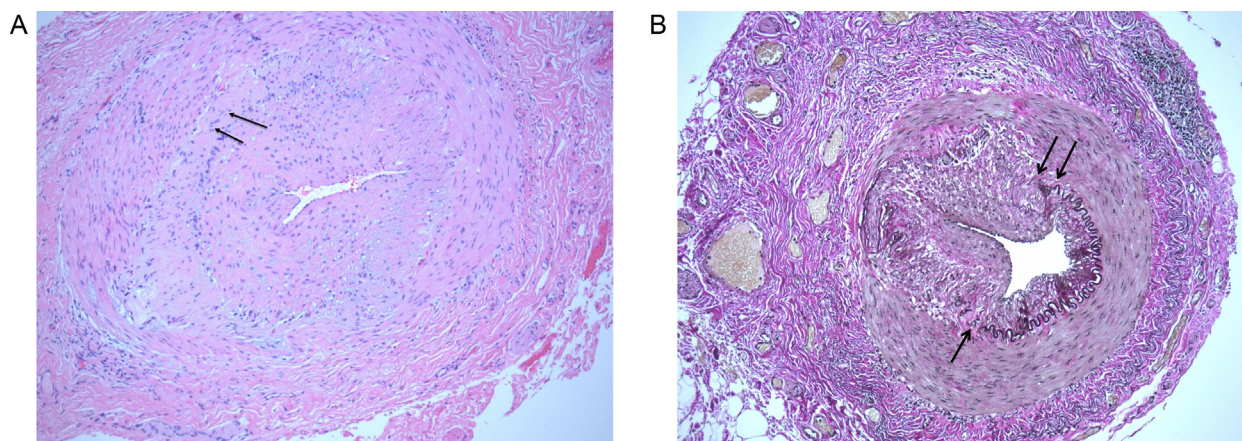


Fig. 2. Digital subtraction angiogram showing (A) focal occlusion of the right axillary artery with reconstitution of the brachial artery and (B) severely stenotic left subclavian and axillary arteries.



**Fig. 3.** Temporal artery biopsy results showing (A) giant cells (black arrows) on H&E stained section, as well as (B) disruption of the internal elastic lamina (black arrows) on a Verhoeff Van Gieson stained section.

Evidence of upper extremity involvement is found in 10–30% of patients [3]. Patients with upper extremity involvement are typically female, younger, and less likely to have headaches, jaw claudication, and anterior ischemic neuropathy [4]. Despite the prevalence of upper extremity involvement in GCA, only 1–2% of patients present with arm claudication or ischemia [5]. Temporary artery biopsy is often negative in patients with large vessel manifestations of GCA [6].

The medical treatment of GCA is well-established; patients are treated with steroids as well as medications to prevent their side effects [1]. Surgical options are less well-defined. There are case reports and small series describing angioplasty for lesions seen in temporal arteritis; there have been conflicting results, with some series reporting success and resolution of symptoms when stenting or angioplasty is used in conjunction with steroids [7–10] and others reporting as many as 50% of patients needing additional procedures [11]. Additionally, one case series on endovascular treatments reports dissection flaps complicating 16 of 40 arteries angioplastied [11]. The data is similarly scant for results with open bypass, again a subject of several case reports [5,9,12–17]. The type of bypass conduit is not always detailed, particularly in those studies published in the rheumatologic literature. Data is therefore, limited to make meaningful conclusions regarding the optimal conduit.

In this patient, we chose to perform a bypass rather than stent the lesions. She had long stenoses on both sides, making endovascular techniques suboptimal. Stenting would also have involved stenting across joints, placing the stents under repetitive stress and possible risk of fracture. A bypass was therefore selected; the choice between prosthetic and vein was made due to her relatively young age, the availability of suitable cephalic vein bilaterally, and the superiority of vein as a conduit in lower extremity bypass.

In the existing literature, steroids are often used in conjunction with surgery [13,14,18]. Here, a steroid taper was started after the second operation because the diagnosis was not definitive until after her second surgery. In some cases, steroids obviate the need for surgery [9,11], but there is not enough literature to determine the best timing of steroid administration.

Renal artery stenosis was found in this patient upon imaging of the rest of her vasculature. Making the diagnosis is important in order to prompt the administration of steroids, but also to initiate evaluation of the rest of the vasculature. The involvement of the aorta and other vessels is frequent in patients with GCA [4]; they have twice the risk of the normal population of having an aortic aneurysm [19].

This patient had an unusual presentation of inflammatory arteritis: she had none of the ACR criteria symptoms and a near-normal ESR. Her arteriogram showed long segment involvement with focal stenosis and dilations, typical of giant cell arteritis. Temporal artery biopsy provided the definitive diagnosis in this patient and should be considered when the appearance of the arterial lesion is unusual, even in the absence of typical GCA features.

#### Conflicts of interest

None of the authors have any personal or financial relationships with other people or organizations that would bias their work.

#### Ethical approval

The patient herself consented to the publication of this case report. As this documents care that was provided and not research performed on the patient, formal ethics committee approval was not obtained.

#### Consent

Written and signed consent for this case report was obtained from the patient. All identifying characteristics were removed as possible while preserving the scientific meaning.

#### Author contribution

Julia Glaser, MD: concept and design, data collection, data interpretation, writing the paper. Rebecca Sharim, MD: design, data interpretation, critical revision of the paper. Belinda Birnbaum, MD: design, data interpretation, critical revision of the paper. Kathleen Montone, MD: concept, data collection, critical revision of the paper. Grace Wang, MD: concept and design, data collection, data interpretation, critical revision of the paper.

#### Guarantor

Grace Wang, MD accepts full responsibility for the work, had access to the data, and controlled the decision to publish.

#### Acknowledgement

The authors have no grants or funding organizations to acknowledge in supporting this research.

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