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



Julia Glaser^{a,*}, Rebecca Sharim^b, Belinda Birnbaum^b, Kathleen Montone^c, Grace Wang^a

- a Division of Vascular and Endovascular Surgery, Department of Surgery, Hospital of the University of Pennsylvania, Philadelphia, PA, USA
- ^b Division of Rheumatology, Department of Medicine, Hospital of the University of Pennsylvania, Philadelphia, PA, USA
- ^c Department of Pathology and Laboratory Medicine, Hospital of the University of Pennsylvania, Philadelphia, PA, USA

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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 vaculitides. 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.

E-mail address: Julia.Glaser@uphs.upenn.edu (J. Glaser).

Her only medications were a lisinopril/hydrocholorthiazide 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 3-4cm 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

^{*} Corresponding author at: Division of Vascular and Endovascular Surgery, Department of Surgery, 3400 Spruce St, 4 Maloney, Hospital of the University of Pennsylvania, Philadelphia, PA, 19104, USA.

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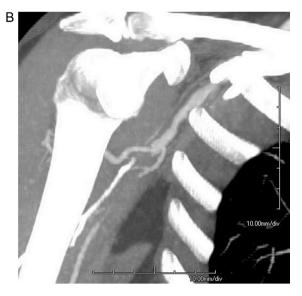


Fig. 1. CT angiogram showing (A) irregularity of the left subclavian and segmental stenoses of the left axillary artery and (B) focal occlusion of the right axillary artery.

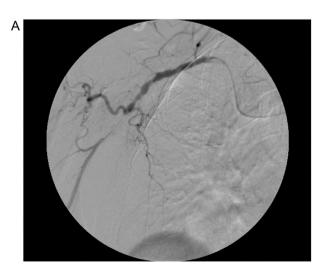
of the vertebral. Her left axillary artery reconstituted inferior to the clavicle.

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.



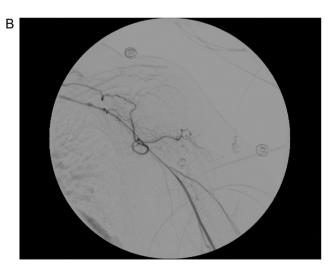


Fig. 2. Digital subtraction angiogram showing (A) focal occlusion of the right axillary artery with reconstitution of the brachial artery and (B) severely stenotic left subclavian and axillary arteries.

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