Rev Port Cardiol. 2016;35(4):237.e1–237.e4

Revista Portuguesa de
Cardiologia
Portuguese Journal of Cardiology
www.revportcardiol.org

CASE REPORT

Acute upper limb ischemia, a rare presentation of giant cell arteritis

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Received 27 April 2015; accepted 13 September 2015
Available online 19 March 2016

KEYWORDS
Giant cell arteritis;
Critical ischemia;
Carotid-humeral bypass

Abstract Giant cell arteritis (GCA) is a systemic large vessel vasculitis, with extracranial arterial involvement described in 10–15% of cases, usually affecting the aorta and its branches. Patients with GCA are more likely to develop aortic aneurysms, but these are rarely present at the time of the diagnosis.

We report the case of an 80-year-old Caucasian woman, who reported proximal muscle pain in the arms with morning stiffness of the shoulders for eight months. In the previous two months, she had developed worsening bilateral arm claudication, severe pain, cold extremities and digital necrosis. She had no palpable radial pulses and no measurable blood pressure.

The patient had normochromic anemia, erythrocyte sedimentation rate of 120 mm/h, and a negative infectious and autoimmune workup. Computed tomography angiography revealed concentric wall thickening of the aorta extending to the aortic arch branches, particularly the subclavian and axillary arteries, which were severely stenotic, with areas of bilateral occlusion and an aneurysm of the ascending aorta (47 mm). Despite corticosteroid therapy there was progression to acute critical ischemia. She accordingly underwent surgical revascularization using a bilateral carotid-humeral bypass. After surgery, corticosteroid therapy was maintained and at six-month follow-up she was clinically stable with reduced inflammatory markers.

GCA, usually a chronic benign vasculitis, presented exceptionally in this case as acute critical upper limb ischemia, resulting from a massive inflammatory process of the subclavian and axillary arteries, treated with salvage surgical revascularization.

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PALAVRAS-CHAVE
Arterite de células gigantes;
Isquemia crítica dos membros superiores;
Bypass carótido-umeral

Isquemia crítica dos membros superiores, uma apresentação rara de arterite de células gigantes

Resumo A arterite de células gigantes (ACG) é uma vasculite de grandes vasos, com envolvimento extracraniano em 10-15% dos casos, afetando preferencialmente a aorta e os seus ramos. Os aneurismas da aorta ascendente são mais frequentes em doentes com ACG, mas são raros no momento do diagnóstico.

Apresentamos o caso de uma doente, caucasiana, com 80 anos de idade, que descrevia dor muscular nos membros superiores, com rigidez matinal dos ombros, desde há oito meses. Nos últimos dois meses referia claudicação intermitente de agravamento progressivo, dor intensa, extremidades frias e o aparecimento de úlceras digitais. Ao exame objetivo não se palpavam pulsos radiais ou cubitais, nem se registavam valores de pressão arterial.

Laboratorialmente, destacava-se anemia normocíticanormocrómica, VS de 120 mm/h e estudo autoimune e infeccioso negativos. A tomografia computorizada com contraste revelou espessamento concêntrico da parede da aorta e dos seus ramos, notoriamente artéria subclávia e axilar, que apresentavam áreas de estenose e oclusão bilaterais e uma dilatação aneurismática da aorta ascendente (47 mm). Apesar da instituição imediata de corticoterapia, verificou-se uma progressão para isquemia crítica aguda dos membros superiores. Consequentemente, foi submetida com sucesso a revascularização cirúrgica através de um *bypass* carótido-umeral. A doente manteve corticoterapia e ao 6.º mês de *follow-up* encontrava-se clinicamente estável, com regressão dos parâmetros inflamatórios.

A ACG é habitualmente descrita como uma patologia indolente e benigna. No entanto, apresentou-se neste caso como uma isquemia crítica aguda dos membros superiores, secundária à intensa resposta inflamatória das artérias subclávias e axilares, obrigando à revascularização cirúrgica.

Introduction

Giant cell arteritis (GCA) is a systemic medium and large vessel vasculitis, with a wide spectrum of presentations, most frequently secondary to involvement of the cranial arteries, the typical symptoms being headache, jaw claudication, visual loss and stroke. Extracranial arterial involvement is described in 10–15% of cases,[1,2] usually presenting as upper or lower extremity claudication, and rarely with acute critical ischemia as the primary finding.

We report a severe and sudden case of acute critical ischemia of the arms with digital necrosis as the presenting features of GCA, successfully treated by salvage revascularization surgery. Only large vessels were involved, mainly the subclavian and axillary arteries, with no involvement of the carotid or cranial arteries.

Case report

An 80-year-old Caucasian woman with a history of hypertension described proximal muscle pain in the arms with morning stiffness of the shoulders and neck, for the past eight months. In the previous two months, she had developed worsening bilateral arm claudication, severe pain with restricted mobility, cold extremities and digital necrosis. She had no palpable radial or cubital pulses and no measurable blood pressure. There were no clinical signs of cranial or lower limb artery involvement and no constitutional symptoms were reported.

Investigation

The patient presented normochromic anemia, elevated erythrocyte sedimentation rate (ESR) (120 mm/h) and C-reactive protein (CRP) (123 mg/l), and negative infectious and autoimmune workup. Contrast computed tomography angiography (CTA) in the arterial phase (Figures 1–3) revealed concentric wall thickening of the aorta extending to the aortic arch branches, particularly the subclavian and axillary arteries, which were severely stenotic with areas of bilateral luminal occlusion, and no involvement of the carotid branches; an aneurysm of the ascending aorta 47 mm in diameter was also detected.

The patient immediately started corticosteroid therapy (prednisone 1 mg/kg daily) and aspirin (100 mg

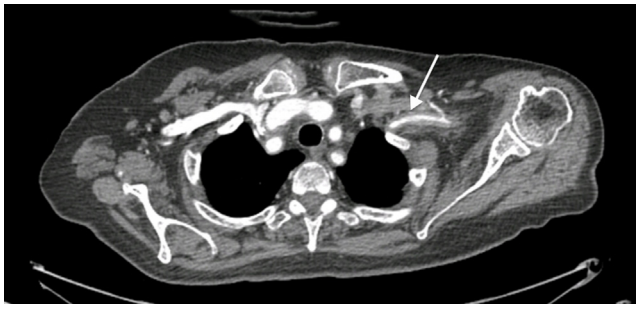

Figure 1 Concentric wall thickening of the left subclavian artery.