Case Report

^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography of giant cell arteritis with lower extremity involvement in association with polymyalgia rheumatica

ABSTRACT

An 80-year-old man presented with new-onset pain in the shoulders and lower extremities and elevated serum inflammatory markers. A clinical diagnosis of polymyalgia rheumatica (PMR) was made, but there was a suboptimal response to glucocorticoid therapy, prompting further evaluation. ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) revealed intense FDG uptake in the arteries of the bilateral lower extremities, head, and neck, but sparing the aorta, suggestive of an uncommon pattern of giant cell arteritis (GCA). There were also imaging signs consistent with PMR, including FDG uptake in the synovium of large joints. This case highlights the uncommon manifestation of GCA with lower extremity involvement and sparing of the aorta. The combination of FDG PET imaging features and elevated serum markers obviated the need for invasive biopsy. One might also conclude that standard FDG PET/CT imaging protocols covering orbits/vertex to thighs incompletely evaluate the extent of arterial distribution of GCA.

Keywords: Diagnosis, diagnostic imaging, giant cell arteritis, polymyalgia rheumatica, rheumatic diseases, vasculitis

INTRODUCTION

This case demonstrates the uncommon presentation of lower extremity giant cell arteritis (GCA) seen on fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) with sparing of the aorta in association with polymyalgia rheumatica (PMR). Symptomatic GCA involving the lower extremity arteries has been sparingly described in clinical rheumatological literature with an overall prevalence of <1% and is considered to be clinically underestimated.[1] To our knowledge, there is only one case describing a similar FDG PET/CT appearance, however, that case was described in the context of generalized vasculitis without specific diagnosis of GCA or PMR.

CASE REPORT

An 80-year-old man presented with a new onset of muscular pain in shoulders and bilateral lower extremities, particularly his thighs and shins, frontal headache, occasional blurring of vision, and fatigue. He denied symptoms of jaw, leg, or arm claudication, fever, and weight loss. Family history was positive for GCA in his mother. Laboratory tests revealed elevated erythrocyte sedimentation rate (ESR) of 77 mm/h (normal, 3–28 mm/h) and serum C-reactive protein (CRP) of 191

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mg/L (normal, 0–8 mg/L). He was placed on 20 mg of prednisone per day with presumptive diagnosis of PMR. Repeat laboratories after a month of steroid therapy showed persistently elevated ESR (73 mm/h) and CRP (127 mg/L). Due to lack of response to steroids, ^{18}F-fluorodeoxyglucose (FDG) PET/CT was performed from vertex to toes on a digital PET/CT scanner (DMI®, GE Healthcare, Milwaukee, Wisconsin, USA) to evaluate for underlying vasculitis [Figures 1 and 2].

The FDG PET/CT showed intense FDG uptake greater than the liver in bilateral lower extremity arteries and their branches, including iliofemoral and tibioperoneal arteries [Figure 1]. Increased uptake was also seen in extracranial head-and-neck arteries including vertebral, carotid, and superficial temporal arteries [Figure 2]. The aorta was normal [Figure 2]. Findings were suggestive of active vasculitis involving large- and medium-sized vessels with sparing of the aorta. FDG PET/CT also showed synovitis and inflammatory type uptake around large joints of the appendicular skeleton, suggestive of PMR [Figure 1]. Antineutrophil cytoplasmic antibodies (c-ANCA and p-ANCA) were negative, and there were no clinical features of small-vessel vasculitis. Based on these findings, a diagnosis of GCA with PMR was made. He was begun on tocilizumab immunosuppressant therapy, and prednisone dose was increased to 60 mg daily. Repeat laboratory tests after a month of therapy showed normalization of ESR (14 mm/h) and CRP (3 mg/L) and significant improvement in previous symptoms such as headache, jaw discomfort, shoulder, and hip stiffness.

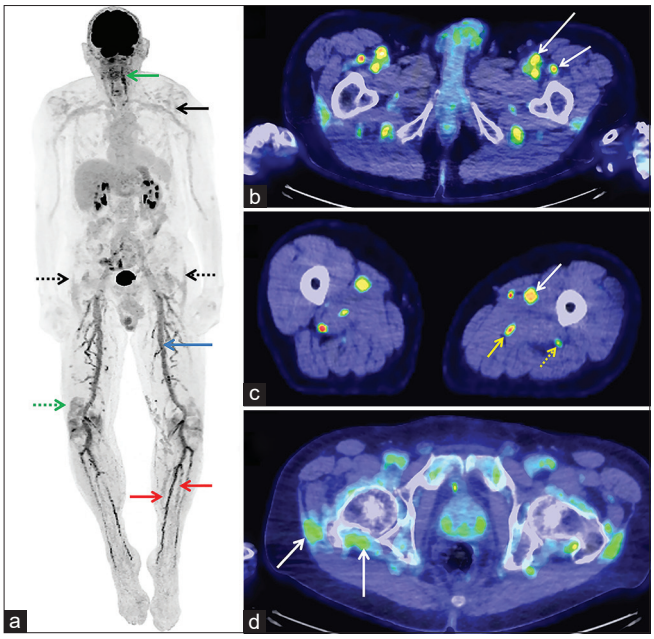

Figure 1: Whole-body maximum intensity projection image (a) shows fluorodeoxyglucose uptake in both iliofemoral (blue arrow) and tibioperoneal (red arrows) and vertebral (green arrow) arteries. Subclavian and axillary arteries show fluorodeoxyglucose uptake less than the liver (black arrow). There is fluorodeoxyglucose-avid synovitis (dotted green arrow) and periarticular uptake (dotted black arrows) suggestive of polymyalgia rheumatica. Axial-fused positron emission tomography/computed tomography images show increased fluorodeoxyglucose uptake in femoral (white arrows, b), anterior tibial (white arrow, c), posterior tibial (solid yellow arrow, c), and peroneal (dotted yellow arrow, c) arteries and periarticular and synovial uptake in hip joints (white arrow, d)

DISCUSSION

GCA is an idiopathic granulomatous medium- and large-vessel vasculitis that typically involves the aorta, extracranial head-and-neck arteries (most common the superficial temporal arteries), and subclavian arteries.[2] GCA with lower extremity involvement is uncommon with only a few small reported case series.[1,3-5] The true prevalence of GCA with lower extremity involvement is likely underestimated clinically as patients can be asymptomatic without claudicatory symptoms.[6] Furthermore, many FDG PET/CT scans performed for vasculitis do not include the legs. GCA with lower extremity involvement typically involves the femorotibial arteries, followed by iliac arteries.[1,3,4,7] GCA with lower extremity involvement may also be seen in patients with PMR.[6] Whole-body FDG PET imaging can play an important role in early detection of lower extremity arterial involvement in asymptomatic patients. Early diagnosis of lower extremity arterial involvement and conveying this information to rheumatologists is important for rapid initiation of immunosuppressant therapy to avoid critical limb ischemia.[8] Whole-body FDG PET provides a one-stop-shop for detection of the extent of active vasculitis as well as concomitant manifestations of PMR.[9] Moreover, the wider availability of digital PET/CT scanners with more sensitive silicon photomultiplier tubes has the potential to further increase the detection of lower extremity arterial involvement. While both visual and semi-quantitative elevations based on maximum standardized uptake value have been described for the evaluation of vasculitis, visual assessment is considered more reliable and reproducible. A visual assessment of FDG uptake greater than the liver has the highest diagnostic accuracy for detection of active vasculitis and monitoring response to therapy.[10] Both FDG uptakes similar to the liver and greater than the liver indicate active vasculitis, while FDG uptake less than the liver or similar to blood pool activity suggests favorable response after therapy on follow-up.[9] The overall sensitivity of FDG PET/CT for diagnosis of vasculitis also increases with elevated serum inflammatory markers and in patients who are glucocorticoid naïve or have glucocorticoid therapy tapered before imaging.[9] Atherosclerotic inflammation is another common cause of FDG uptake in lower extremities but is typically patchy with uptake less than liver, unlike the segmental uptake in GCA.[9] Takayasu arteritis is another large-vessel arteritis but is seen in middle-aged women <50 years, with involvement of aorta and great vessels.[9] The combination of

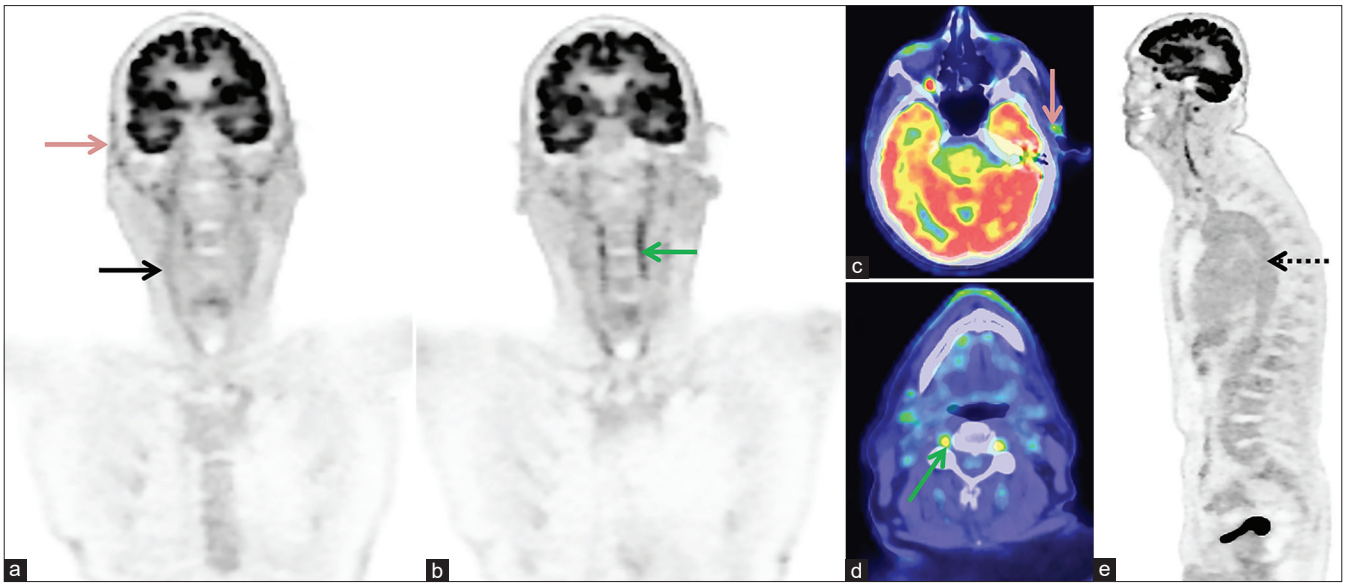

Figure 2: Coronal positron emission tomography only images anteriorly (a) and posteriorly (b) and axial-fused positron emission tomography/computed tomography images of the neck (c) and head (d) show increased fluorodeoxyglucose uptake in external carotid arteries (solid black arrow, a), bilateral superficial temporal arteries (pink arrows, a and c), and vertebral arteries (green arrows, b and d). The aorta is normal with fluorodeoxyglucose uptake within the range of normal for a modern silicon photomultiplier digital positron emission tomography scanner (dotted black arrow, e)

clinical history (age >50 years) PMR, elevated serum markers, and superficial temporal artery involvement was quite diagnostic of GCA in this case despite the sparing of aorta and lower extremity involvement.[2] Thus, recognition of FDG PET imaging features of GCA and PMR can obviate the need for invasive biopsy and guide appropriate immunotherapy.

Exception for five authors

We sincerely request five authors to be listed for this submission as the patient was jointly managed by two rheumatologists, and it was crucial to have both of their inputs on the case. They provided the necessary case history and the clinical context for the discussion. All five authors contributed equally to the manuscript writing and approved the final draft.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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