Giant Cell Arteritis Presenting as PUO

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Abstract
Giant cell arteritis (GCA) is a primary granulomatous vasculitis affecting the large and medium sized arteries. We present here a case of GCA with pyrexia of unknown origin (PUO) as the presenting manifestation in the absence of other typical features. On evaluation, the patient had raised inflammatory markers with features of large vessel vasculitis on whole body PET-CT scan. The colour doppler ultrasonography (CDUS) of the temporal arteries showed bilateral halo sign. Since bilateral ‘halo sign’ is more specific for the diagnosis of GCA, temporal artery biopsy is not mandatory. If CDUS is not conclusive, then biopsy can be considered being an invasive procedure.

Introduction
Giant cell arteritis (GCA) also referred to as ‘temporal arteritis’, is a primary granulomatous vasculitis affecting the large and medium sized arteries. It usually affects adults older than 50 years. The common symptoms are headache, fever, weight loss, jaw claudication and visual symptoms associated with elevated inflammatory markers. Here, we report a case of GCA with pyrexia of unknown origin (PUO) as the initial presenting manifestation and in the absence of other typical manifestations.

Case Report
A 61 year old male presented with high grade fever and chills of 1 month duration. He had no associated cough, dysuria, rashes, joint pain, abdominal pain, malena or altered bowel habits. He had no recent travel history. He had reduced appetite, but there was no significant weight loss. He had no comorbidities or history of tuberculosis in the past. He was a smoker and had stopped smoking since 1 year. On examination, he had bilateral axillary nodes of 2 X 2 cm size. Otherwise, all his system examinations were within normal limits. He had a hemoglobin of 10.3 g/dl, total leukocyte counts of 8820 cells/cu.mm, platelets of 2,12,000 cells/cu.mm and ESR of 54 mm/hour and CRP of 185.1 mg/L. His peripheral smear revealed normocytic normochromic anemia, but had no abnormal cells or parasites. His Chest X ray and urine routine was normal. All infectious disease work-up including culture, serologies were negative. Computed Tomography (CT) of the chest and abdomen were normal. Excision biopsy of axillary node was suggestive of reactive lymphoid hyperplasia. As a part of PUO workup, PET-CT scan of the whole body was done. On PET-CT scan there was symmetrical linear metabolic activity associated with peri-arterial fat stranding in bilateral axillary, subclavian, carotid, aortic arch, thoracic aorta, descending aorta and bilateral iliac arteries suggestive of a large vessel vasculitis (Figure 1). In view of his age, a diagnosis of giant cell arteritis was considered and colour doppler ultrasonography (CDUS) of both the superficial temporal arteries were done. The temporal arteries had an increased diameter with ‘halo sign’ (Figure 2) present on both sides (Maximum thickness of halo was 1.4 mm on the right and 0.9 mm on the left side). Hence, the diagnosis of GCA was confirmed. Since the presence of bilateral ‘halo signs’ is more specific for diagnosis of GCA, temporal artery biopsy was not done. The patient was started on oral prednisolone at a dose of 1 mg/kg with calcium and vitamin D supplements and patient improved symptomatically. To our knowledge, very few cases of GCA with PUO as the presenting manifestation has been reported in literature.2,3

Discussion
GCA is a chronic granulomatous vasculitis with a special predilection for extracranial branches of carotid arteries.¹ GCA is rare in Asian population, with only small case series and isolated case reports published from India.³,4 Most of the cases reported from India were in the age group of

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CDUS is an excellent non invasive tool to assess the temporal arteries. The presence of a dark hypoechoic area (due to vessel wall oedema and thickening) around the temporal artery is referred to as ‘halo sign’. The hypoechoic area should be circumferential and has to be demonstrated in both longitudinal and cross sectional views. Other than halo sign, the presence of stenoses and occlusion can be demonstrated in temporal arteries. The presence of unilateral ‘halo sign’ has a sensitivity of 82% and specificity of 91%. If it is present on both sides, then the specificity increases to 100%. Similarly in our patient, the demonstration of bilateral halo sign confirmed the diagnosis of GCA and hence there was no need for a temporal artery biopsy. We should remember that CDUS is an observer dependent test and involvement of temporal artery can also be found in other conditions like Behçet syndrome, polyarteritis nodosa, ANCA (Anti neutrophil cytoplasmic antibody) associated vasculitis,10,11

The typical findings of large vessel vasculitis in a MRI are wall thickening and increased mural gadolinium contrast enhancement. It has a sensitivity of 81% and specificity of 97%. PET-CT scan of whole body can be considered in patients with fever and inflammation of unknown origin. It can be considered in patients with suspected large vessel vasculitis with negative biopsy, absent halo sign on CDUS and absent wall thickening on MRI, as it can detect early inflammation. In atherosclerosis there will be an increased uptake, but it will be patchy and of less intensity in comparison to the diffuse and high intensity uptake in LVV. PET-CT also helps in the detection of features suggestive of an associated Polymyalgia rheumatica.12

The gold standard test for diagnosis of GCA is temporal artery biopsy. It is an invasive procedure and it can be false negative due to the presence of skip lesions. In patients with palpable abnormalities of the vessel, smaller segment (1-2cms) may be excised. But in others, a larger section (4-6cms) of the vessel has to be excised and multiple sections need to be studied by the histopathologist.1

The initial treatment is oral steroids at a dose of 1mg/kg/day for a period of 4 weeks. Then, the steroid dose has to be gradually tapered every week or every 2 weeks by 10% till a dose of 20 mg and after that even slowly.1 Intravenous methylprednisolone can be given as a pulse therapy in patients with visual complications. On a long term, drugs like methotrexate, tocilizumab, abatacept, ustekinumab may be considered as steroid sparing agents. Since patients might be on long term steroids, measures to prevent side effects like osteoporosis must be initiated early.13

Conclusion
In the algorithm for diagnosis of GCA, clinical examination and inflammatory markers can be followed by CDUS of both the temporal arteries. It is a simple non invasive tool compared to the invasive temporal artery biopsy. If CDUS demonstrates ‘halo sign’ in bilateral temporal arteries, then the diagnosis is confirmed without need for a biopsy. If CDUS is not conclusive, then temporal artery biopsy can be done. If the biopsy is also negative, then PET-CT of whole body can be done on the background of a strong clinical suspicion.10

References