Pyrexia of Unknown Origin- An elusive diagnosis

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Abstract

Giant cell arteritis (GCA) is a systemic inflammatory disorder that affects large vessels. Here we present the case of a 59 year old male who presented to us with complaints of fever and generalised tiredness of 6 weeks duration. He had received 2 courses of antibiotics with no respite. Investigations from outside showed elevated erythrocyte sedimentation rate (ESR) and prostate specific antigen (PSA). He was evaluated in detail as FUO. His fever persisted despite maximal dose of antipyretics, leading to high suspicion of malignancy. However, investigations were not suggestive of any malignancy. On day 3 of admission, he complained of pain on mastication. Examination revealed prominent temporal arteries, with normal pulsations without any tenderness. Fundus examination was normal. With high index of suspicion, temporal artery was biopsied and was diagnostic of giant cell arteritis. Though GCA classically presents with headache, visual changes, masticatory claudication, and symptoms of polymyalgia rheumatica, on rare occasion, GCA may present with fever as the only dominant symptom. GCA without headache is an uncommon presentation, which occurs in nearly 20% of patients.

Keywords: Giant cell arteritis; Fever; Headache

Case Report

A 59 year old gentleman presented with 1 month history of fever which was high grade and intermittent associated with myalgia, tiredness and weight loss with no headache, arthralgia or visual disturbances. There was no history of systemic symptoms, comorbid illnesses, smoking or alcohol intake. He had consulted his family physician, screening for tropical diseases was negative and had received 2 courses of antibiotics over the past one month but symptoms persisted.

On examination, temperature was 102°F, pulse rate was 106 beats per minute and blood pressure was 120/70 mmHg. His right temporal artery was mildly prominent but was pulsatile, non-tender and was not thickened. System examination was unremarkable except for a mildly enlarged nodular prostate on per rectal examination. Initial investigations showed Hemoglobin of 13.2 g/dL, total count of 6790/mm3 with 68% neutrophils and an ESR of 135 mm/hour. His urine microscopy was normal; so were his blood picture, chest X-ray and per rectal ultrasonography, both of which were less in favour of a neoplasm. By day 4, he complained of neck pain while chewing, which was not there previously. With high clinical suspicion of GCA, in view of a prominent right temporal artery which was pulsatile and non-tender and a possible symptom of jaw claudication; temporal artery biopsy was performed – 2 cm of the right frontal branch was obtained for histopathology which was diagnostic of GCA (Figure 1).

Thus he satisfied the ACR criteria for GCA and was started on oral steroids, with calcium and vitamin D supplementation, with which he improved dramatically; his ESR dropped to 6 mm/hour within a month of initiating therapy. Ophthalmology evaluation was done prior to treatment and was normal. Angiography was done to rule out involvement of vertebral vessels which was
normal. His steroid dose is being tapered at present and he is stable at the end of one year of follow-up.

**Figure 1:** Biopsy specimen of the patient’s right temporal artery.

**Figure 2:** Halo sign in duplex ultrasonography.

**GCA** is considered a medical emergency due to the complication of acute loss of vision. Other early complications include stroke and late complications include aortic aneurysm and dissection [9]. Management includes oral steroids for a minimum duration of 2 years and low dose aspirin for ischemia prevention. Early visual loss is managed with pulse dose of intravenous methyl prednisolone [10,11]. Initiation of the treatment with steroids should not be delayed for temporal artery biopsy as delay in treatment can lead to visual loss. Treatment with adjuvant therapies like tocilizumab, methotrexate etc should be considered if there is relapse on steroid therapy or resistant disease [12]. Magnetic resonance angiogram of other arterial systems should be done as other large arteries can also be affected. The traditional concept of GCA focused on cranial symptoms of headache and visual loss is gradually changing and more attention is given to the constitutional symptoms [4].

**Conclusion**

- One in Ten cases of GCA can present as pyrexia of unknown origin; hence it is an important differential diagnosis in pyrexia of unknown origin in the elderly.
- Though usually low grade, 15% patients can develop fever >102°F
- 20% of cases may present without headache.

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