Letter to Editor

Temporal arteritis with normal erythrocyte sedimentation rate and C-reactive protein at presentation

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Dear Editor,

Temporal arteritis or giant cell arteritis (GCA) is an inflammatory vasculopathy involving medium and large arteries with a predilection to affect the temporal arteries and other branches of the external carotid artery. It preferentially affects arteries with internal elastic lamina. It is the most common form of vasculitis in the white population over the age of 50. Rapid diagnosis and management are of paramount importance in GCA due to its potential to cause irreversible vision loss. It's a do not miss diagnosis and prompt diagnosis can avert visual loss. Even though temporal artery biopsy (TAB) remains the gold standard, GCA can be diagnosed by clinical criteria. Erythrocyte sedimentation rate (ESR) >50 is a major criterion for diagnosis [1]. C-reactive protein (CRP) has better sensitivity than ESR for diagnosis of GCA. It is very rare to have normal ESR and CRP in GCA. GCA patients with normal ESR and CRP have fewer systemic symptoms and more visual symptoms. If clinical suspicion is strong you have to do TAB even if ESR and CRP are normal.

A 68-year-old man presented with a 1-week history of the left-sided headache and facial pain. The headache was moderate in intensity and continuous non-throbbing without any vomiting. He also complained of jaw claudication. He also reported episodes of binocular diplopia without any diurnal variation. There was no history of any blurring of vision. There was no history of fever, neck, or shoulder pain. There was no history of arthritis, arthralgia, hypertension, diabetes mellitus, or oral ulcers. There was no history of any headache in the past or recent varicella zoster infection.

His blood pressure was 130/80 mm of Hg and his pulse rate was 68/min. He was afebrile. Examination showed normal fundi and extraocular movements were normal. There were no focal neurological deficits. The left temporal artery was thickened, tender, and distal pulsation was absent. There was no scalp tenderness. Investigations showed an ESR of 18 mm/h and CRP of 5.15 mg/L (normal <6 mg/L). Other investigations are shown in Table 1.

He was started on prednisone 40 mg/day. He subsequently underwent a magnetic resonance imaging (MRI) brain which was normal. Doppler study of the temporal artery showed reduced peak systolic velocity (PSV) from the left temporal artery (28 cm/s). It was thickened, band-like, and non-compressible. There was no halo sign. Right temporal PSV was 50 cm/s. Left TAB (1 cm) was done 2 weeks after the presentation. ESR and CRP done on the day of the biopsy were 57 mm/h and 23.31 mg/L, respectively. The biopsy section showed an artery with a blood clot within the lumen. There is a thickening of the intimal layer with moderate inflammation of the vessel wall involving the circumferentially tunica media and adventitia. The inflammation is composed of lymphocytes, macrophages, plasma cells, and occasionally giant cells (Fig. 1). He was treated with prednisolone 40 mg/day and aspirin and his headache subsided in a week and jaw claudication subsided in a month.

GCA is the most common vasculitis in the elderly with an incidence of 15–30/100,000 persons aged >50 years in the North American and European countries. However, the incidence is believed to be much lower in Asian countries, with the incidence in Japan reported to be as low as 1.47/100,000. In Europe and North America, the female-to-male ratio is nearly 3:1, whereas, the ratio tends to be 1:1 in Western Asia. The clinical presentation of GCA in India has no gender preference and occurs in a slightly younger age group. The pathogenesis of GCA is complex and includes a dysregulated immune response that affects innate and adaptive immunity. GCA is a disease of the elderly and is almost exclusively seen in individuals over the age of 50 years old. Clinical features include headache, jaw claudication, and visual symptoms in the form of amaurosis fugax or diplopia. Systemic symptoms such as fever, fatigue, and weight loss are common. Approximately, 50% have associated polymyalgia rheumatica. Although permanent vision loss is considered to be the most feared complication of GCA, the main factors responsible for mortality in this pathology are cerebral ischemia and aortic aneurysms and dissection. High-resolution MRI and magnetic resonance angiography, FDG PET, and color Doppler can show findings relevant to the diagnosis of GCA. In centers with appropriate radiology expertise, a European rheumatology consensus guideline has proposed Doppler ultrasound as the first-line confirmatory test for GCA in lieu of TAB. Sometimes, accidentally detected halo signs at the level of vertebral arteries during a routine cervical ultrasound may be the

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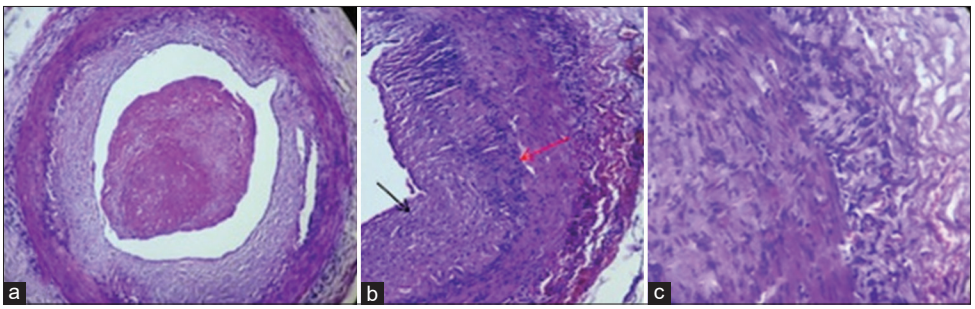

Figure 1: (a) Low magnification showing a cross-section of the artery with central thrombosis; (b) inflammatory cells in concentric pattern (red arrow) and thickening of intimal layer (black arrow); and (c) high magnification showing chronic inflammatory cells in tunica media and adventitia

Table 1: Blood investigations of the patient

Hemoglobin	14.3 g/dL
Hematocrit	43%
WBC count	9420 cells/mm^3
Neutrophils	69%
Lymphocytes	30%
Eosinophils	01%
Platelets	4.22 lakhs/mm^3
ESR	18 mm/h
CRP	5.15 mg/L
Total bilirubin	0.41 mg/dL
Direct bilirubin	0.09 mg/dL
SGOT	57 U/L
SGPT	48 U/L
Alkaline phosphatase	69 U/L
Total protein	6.5 g/dL
Albumin	3.4 gm/dl
Blood glucose	116 mg%

WBC: White blood cell, ESR: Erythrocyte sedimentation rate, CRP: C-reactive protein, SGOT: Serum glutamic-oxaloacetic transaminase SGPT: Serum glutamic pyruvic transaminase

first clue for GCA in stroke. ESR and CRP may be normal in 4% of TAB-proven GCA patients [2]. One patient out of 17 patients in the Indian series had normal ESR and CRP was normal in three out of 11 patients [3,4]. The degree of ESR elevation is correlated with the degree of anemia. In one study, all patients with hematocrit >40% had ESR <50 whereas all 18 patients with hematocrit <40% had an ESR of more than 50 mm/h [5]. An ESR of 30 mm/h or more is more sensitive in detecting GCA [6].

Laboratory investigations usually reveal elevated ESR, CRP, and plasma viscosity. Anemia, generally normocytic normochromic, leukocytosis, thrombocytosis, elevated plasma fibrinogen, and decreased serum albumin are often seen. Liver enzymes may be elevated. TAB is diagnostic. An algorithm without biopsy has been suggested based on color Doppler [7].

Historically, ESR has been considered one of the most useful markers to predict the likelihood of having GCA. A normal ESR makes GCA unlikely but does not rule it out. Approximately, 90% of biopsy-confirmed GCA patients have ESR >50 mm/h. Only 3.6% have ESR <30 mm/h [4]. CRP levels may be more sensitive in making the diagnosis. CRP was 100% sensitive and ESR was 92% sensitive [4]. Other laboratory abnormalities including anemia and thrombocytosis were less prevalent in those with normal ESR/CRP. In our case, there was thrombocytosis. In patients suspected of having GCA, an elevated platelet count >4,00,000/mm^3 is a useful marker of positive TAB [8]. At 2 months follow-up, our patient's ESR and CRP were 20 mm/h and 6 mg/L, respectively. His steroid dose was reduced to 10 mg/day and was started on azathioprine 50 mg twice daily. At 6 month follow-up, he is asymptomatic.

In conclusion, ESR and CRP can be normal early in the course of GCA and should be repeated if clinical suspicion is strong. In such cases, TAB should be done and can be positive even after 2 weeks of steroid therapy.

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