Temporal Arteritis in a 41 Year Old Male: A Case Report and Review of Literature

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Abstract

Background: Giant cell arteritis (GCA) or temporal arteritis (TA) is a necrotizing medium to large vessel arteritis of unknown etiology that was described in 1932 by Horton. It commonly afflicts elderly people and may present with diverse manifestations such as severe headache, impairment of vision, polymyalgia rheumatica, tenderness of the temporal arteries, a highly elevated erythrocyte sedimentation rate (ESR) and a characteristic abnormal temporal artery biopsy. It is rare in individuals less than 50 yrs old. Age of more than 50 yrs old is the first of five clinical criteria formulated by the American College of Rheumatology. In the largest series reported, all patients were ≥50 years, and 95% of them were >60 years. In the last three decades, only 13 patients <40 years old have been reported with TA. We report here a case of a relatively young patient presenting with clinical features of TA with a negative temporal artery biopsy.

Setting: St. Luke’s Medical Center- a tertiary care hospital

Keywords: Giant cell arteritis, temporal arteritis

Case Report: Patient is a 41 y/o male who presented with a 3 week history of severe left sided frontal headache with intermittent episode of moderate grade fever, blurring of vision, diplopia on the left eye and left jaw claudication. This was followed by an onset of moderate pain over the right shoulder with limited range of motion. Pertinent physical examination revealed a pulsating, warm and tender to touch left temporal artery, with scalp tenderness. The right temporal artery was essentially normal. There was no Brundzinksi’s and Kernig’s signs. Other neurologic examination were unremarkable. Laboratory investigations revealed: Complete blood count (CBC) Hgb 11.6 g/dl, Hct 33.5%, RBC 4.06 mil/mm3, white blood count at 6,240 mm3, neutrophils 68%, lymphocytes 22%,monocytes 9% and platelet count of 144,000/mm3 , ESR 90 mm/hr., CRP 34.9 mg/dl. Brain CT scan showed normal results as well as cerebrospinal fluid analysis. A left temporal artery biopsy with length of 2cm was obtained. It was grossly palpably normal but histopathology revealed intimal fibrosis of the tunica media devoid of any inflammatory infiltrates. He was managed as a case of temporal arteritis and was started on prednisone at 1mg/kg/day with remarkable improvement and had follow-up 3 months later free of complications on tapering dose of corticosteroids.

Introduction

Giant cell arteritis (GCA), also known as temporal arteritis, cranial arteritis or Horton’s disease, is the most common form of systemic necrotizing vasculitis in individuals who are over 50 years of age.1 Its organ involvement may be widespread and the spectrum of clinical manifestations are principally related to ischemic cranial involvement such as headaches, jaw claudication, scalp tenderness and transient or permanent visual symptoms. It may be associated with constitutional symptoms such as fever, weight loss, anorexia or fatigue.2,3,4 Characteristic histological manifestation is a granulomatous inflammation involving the large and medium sized arteries.5,6 Delayed treatment may result to devastating complications such blindness, stroke, and aortic aneurysms.

Discussion

This case highlights the challenges in the diagnosis of temporal arteritis in a relatively young 41 yr old male patient. The criteria set by the American College of Rheumatology for the formal classification of the condition include age >50 years, recent localized headache, temporal artery tenderness, ESR >50 mm/hour, and a positive temporal artery biopsy. The presence of three or more of these criteria is associated with more than 90% sensitivity and specificity for the diagnosis of the disease. The age criterion is of special importance, as this disease is almost exclusively seen in patients over 50 years old.6,7 In the largest series reported, all patients were ≥50 years, and 95% of them were >60 years. The incidence increases dramatically with age, rising by almost tenfold in the ninth decade.8 In the last three decades, only 13 patients <40 years old have been reported with TA.9,10

The level of clinical suspicion for GCA should be based on patient age, clinical symptoms, laboratory evaluation, and imaging findings. The wide spectrum of clinical manifestations can be divided into those related...
to tissue ischemia from vascular lesions and those related to a systemic inflammatory response. Almost any large or medium-sized artery in the body may be involved, including those of the limbs, liver, intestine, lungs, uterus, breast and skin.\(^6,7,11\) Polymyalgia rheumatica is accepted as part of the clinical spectrum of GCA, occurring concomitantly with temporal arteritis in up to 50% of patients.\(^4\) An elevated ESR (more than 50 mm/1st hour) is a common laboratory finding; however, up to 5% to 10.8% of patients may have an ESR lower than 50 mm/hr.\(^12,13,14\) Other laboratory tests include elevated CRP, fibrinogen and ALP, anemia and leukocytosis.\(^6,7\)

In view of these variable clinical presentations of GCA, the value of pathological confirmation becomes important especially in atypical cases. The presence of jaw claudication, diplopia and temporal artery abnormalities correlates with a high probability of a positive histology. GCA is characterized by the discontinuity of vascular inflammation, hence, early biopsy with long arterial specimens (longer than 20mm) showing palpable tender lesions are needed to yield the positive results.\(^15\) The positive rate for temporal artery biopsy has been reported to range from 75% to 96%.\(^16,17,18\) with characteristic pathological features showing the presence of giant cells, mononuclear infiltrates or granulomas in association with the different features of necrotizing arteritis, and rarely, fibrinoid necrosis.\(^6,19\) Other recognized pathological findings are those of healed arteritis, and are distinguished from changes seen in arteriosclerosis by the presence of focal mononuclear aggregates, and long breaks in elastic lamina.\(^19,20\) Since arterial wall inflammation is segmental, the 2 temporal arteries may also be unevenly involved. Therefore, histological signs of inflammation may be missed in temporal artery biopsy (TAB) performed in arteritis-free segments. As a result, in most studies 10%–20% of TAB are reported as negative in patients with GCA, although the rate may be as high as 40%.\(^19,20\) The study by Breuer\(^21\) suggest that performing bilateral temporal artery biopsies increases the diagnostic sensitivity of the procedure by up to 12.7% compared to unilateral biopsies, which was likewise observed in a study by Khalifa\(^22\), wherein the rate of biopsy positive GCA increased to 73% from 64.6% when a contralateral biopsy was performed.

Our patient’s unilateral TAB of 2cm length showed intimal fibrosis of the tunica media devoid of any inflammatory infiltrate. This finding was also seen in 27% of cases in a series of histopathologic findings of temporal arteritis.\(^22\) A case by Petzold\(^23\) also described a patient presenting with symptoms of GCA but with histopathologic features of a rapidly progressive intimal fibrosis without evidence of inflammation. On the other hand, a normal biopsy can be missed in temporal artery biopsy (TAB) performed in patients with GCA, the value of pathological confirmation becomes important especially in atypical cases. The presence of jaw claudication, diplopia and temporal artery abnormalities correlates with a high probability of a positive histology. GCA is characterized by the discontinuity of vascular inflammation, hence, early biopsy with long arterial specimens (longer than 20mm) showing palpable tender lesions are needed to yield the positive results.\(^15\) The positive rate for temporal artery biopsy has been reported to range from 75% to 96%.\(^16,17,18\) with characteristic pathological features showing the presence of giant cells, mononuclear infiltrates or granulomas in association with the different features of necrotizing arteritis, and rarely, fibrinoid necrosis.\(^6,19\) Other recognized pathological findings are those of healed arteritis, and are distinguished from changes seen in arteriosclerosis by the presence of focal mononuclear aggregates, and long breaks in elastic lamina.\(^19,20\) Since arterial wall inflammation is segmental, the 2 temporal arteries may also be unevenly involved. Therefore, histological signs of inflammation may be missed in temporal artery biopsy (TAB) performed in arteritis-free segments. As a result, in most studies 10%–20% of TAB are reported as negative in patients with GCA, although the rate may be as high as 40%.\(^19,20\) The study by Breuer\(^21\) suggest that performing bilateral temporal artery biopsies increases the diagnostic sensitivity of the procedure by up to 12.7% compared to unilateral biopsies, which was likewise observed in a study by Khalifa\(^22\), wherein the rate of biopsy positive GCA increased to 73% from 64.6% when a contralateral biopsy was performed.

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Our patient’s unilateral TAB of 2cm length showed intimal fibrosis of the tunica media devoid of any inflammatory infiltrate. This finding was also seen in 27% of cases in a series of histopathologic findings of temporal arteritis.\(^22\) A case by Petzold\(^23\) also described a patient presenting with symptoms of GCA but with histopathologic features of a rapidly progressive intimal fibrosis without evidence of inflammation. On the other hand, a normal biopsy can reach 40% to 80% in patients treated with steroids for more than one week\(^24,25\) while some can remain positive for up to 4–6 weeks after commencing high dose corticosteroid.\(^26,27\) Our patient only received 2 days of high dose corticosteroid before TAB. It is worth stressing that a negative TAB does not exclude the diagnosis of GCA, if clinical suspicion for GCA is high and biopsies are negative, several authors\(^28,29,30\) suggest pursuing imaging studies such as ultrason, CT or MR angiography of the aorta and great vessels. Although this was not performed in our case, ultrasonographic changes would show the presence of a dark halo around the arterial lumen, edematous wall stenoses or occlusions.\(^28\) Both CT and MRI should be reserved for patients who are suspected of having large artery involvement.

Once the diagnosis of GCA is seriously considered, treatment with steroids should be instituted at once in order to prevent serious neuro-ophthalmic complications.\(^31\) which can happen early in the course of the disease; with permanent vision loss occurring in up to 20% of patients. The optimum dose and duration of prednisolone is still controversial.\(^32,33,34,35\) It can extend from 20 to 80 mg per day, with a maintenance dose of 20 mg for about two year.\(^36,37\) A gradual steroid taper is considered when clinical symptoms abate and laboratory markers normalize. Most patients can discontinue steroids after 1–2 years of treatment. The median time to steroid discontinuation was 21.6 months.\(^38\)

**Conclusion**

Diagnosis of temporal arteritis is often delayed due to poor recognition of early, often non-specific symptoms. Hence, it should be considered in all patients (even in those <50 years) presenting with temporal artery related symptoms or signs, since it can potentially cause devastating neuro-ophthalmic complications once treatment is delayed.

**References**


