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Giant cell arteritis with internal carotid artery stenosis and third nerve palsy: a case report

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ABSTRACT

Background:

Giant cell arteritis or temporal arteritis is a necrotizing medium to large vessel arteritis of unknown etiology that was firstdescribed in 1932 by Horton. It commonly afflicts elderly people and may present with diverse manifestations such as severe headache, impairment of vision,ophtalmoplegia, polymyalgia rheumatica, tenderness of the temporal arteries, a highly elevated erythrocyte sedimentation rate (ESR) and a characteristic abnormal temporal artery biopsy.

Case report:

We report a 67-year-old man with new onset temporal headache, moderately elevated ESR, cranial nerves palsy, and high grade internal carotid artery stenosis, who was finally diagnosed as giant cell arteritis, although temporal artery biopsy was normal.

Discussion:

We conclude that, giant cell arteritis should be considered in all patients (even in those with ESR <50 and negative biopsy) presenting with third nerve palsy and temporal artery related symptoms or signs.

KEYWORDS : Gaint cell arteritis, Cranial nerves palsy, Carotid Stenosis,temporal arteritis

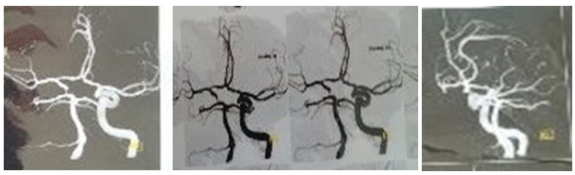
INTRODUCTION

Giant cell arteritis (GCA), also known as temporal arteritis (TA), cranial arteritis or Horton's disease, is the most common form of systemic necrotizing vasculitis in patients more than 50[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, transient or permanent visual symptoms and ophtalmoplegia. It may be associated with constitutional symptoms such as fever, weight loss, anorexia or fatigue[2,] .Characteristic histological manifestation is a granulomatous inflammation involving the large and medium sized arteries[2].Delayed treatment mayresult indevastating complications such as blindness, stroke, and aortic aneurysms.We report a patient presenting with third nerve palsy and visual loss from TA, with negative temporal artery biopsy and high grade stenosis of internal carotid artery.

CASE RESENTATION

A 67-year-old man was admitted to the neurology ward of Rasool Akram Hospital affiliated to Iran University of Medical Sciences, in August 2014 with a 10-day history of severe rightsided frontotemporal headache,right eye blurred vision, right eye ptosis, and jaw claudication.He also reported a moderate pain over his limbs and shoulders. He was a smoker and his past medical history was negative for diabetes mellitus, hypertension,and dyslipidemia . On examination, there was no tenderness over temporal arteries and their pulses were normal. There wasright eye partial ptosis and pallor of right optic disc. Right pupil was sluggish in response to light and visual acuity ofright eye was just light perception.Examination of right ocular movements revealed a deficit in adduction, supraduction and infraduction. Evaluation of left eye and other neurologic examinations were unremarkable. Laboratory investigations revealed: WBC=8400, Hb=16.4, HCT=47.5, platelet count=323000, erythrocyte sedimentation rate (ESR) =45 and c-reactive protein (CRP) =24mg/L. Spiral braincomputed tomography(CT) scan and brain magnetic resonance imaging(MRI) were normal.Nonetheless, brain magnetic resonance angiography (MRA) and ultrasound of carotid arteries demonstrated a significant stenosis in right internal carotid artery (ICA) and halo sign (figure 1). Prednisolon 60 mg daily was started and temporal artery biopsy was done after one week, which was negative. Brain conventional angiography also showed high grade stenosis in right ICA and there was no evidence of aneurysm. He was managed as a case of temporal arteritis and inflammatory markers(ESR and CRP) returned to normalvalues. Besides theimprovement in ptosis and ocular movements, there was no significant change in visual acuity.After four weeks, we tapered prednisolon to confirm the diagnosis of GCA. ESR and CRPraised again; therefore, we resumed steroid therapy.

Figure 1.Brain magnetic resonance angiography showing high grade stenosis of right internal carotid.



DISCUSSION

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 forthe diagnosis of the disease[3]. The level of clinical suspicion for GCA should bebased on patient age, clinical symptoms, laboratoryevaluation, and imaging findings. Our patient had an acute onset temporl headache, third nerve palsy, visual loss and elevated ESR and CRP. Temporal arteritis can present with acute painful third nerve palsy that often improves rapidly with steroid treatment [4]. Also it is important to recognize that 21.2% of patients with GCA and visual loss and ocular symptoms do not have any systemic symptoms of GCA[5]. Thus in persons older than 55 years, developinganterior ischemic optic neuropathy (AION), acute third nerve palsy and abnormal CRP with or without elevated ESR should raise the suspicion for gaint cell arteritis[4, 5]. The wide spectrum ofclinical manifestations can be divided into those related to tissue ischemia from vascular lesions and those relatedto a systemic inflammatory response. Almost any large ormedium-sized artery in the body may be involved, includingthose of the limbs, liver, intestine, lungs, uterus, breast andskin[3, 6]. Polymyalgia rheumatica is accepted as part of theclinical spectrum of GCA, occurring concomitantly withtemporal arteritis in up to 50% of patients, as occurred in our patient. 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[7]. Other laboratory tests include elevated CRP, fibrinogen and ALP, anemia and leukocytosis[3, 4]. The study by Von Blotzeim suggested that ESR is normal in 15% of GCA patients and GCA with normal ESR is not rare; hence, such a patient should be investigated with other blood studies. However, a retrospective study showed although most patients with GCA have both an elevated ESR and CRP, normal ESR and elevated CRP is also consistent with GCA . Our patient had an ESR lower than 50, but an elevated CRP. In view of these variablepresentations ofGCA, the value of pathological confirmation becomesimportant especially in atypical cases. The presence of jawclaudication, diplopia and temporal artery abnormalitiescorrelates with a high probability of a positive histology. GCAis characterized by discontinuity of vascular inflammation; hence, early biopsy with long arterial specimens (longerthan20mm) showing palpable tender lesions is needed to yieldthe positive results. The positive rate for temporal arterybiopsy has been reported to range from 75% to 96%[8], withcharacteristic pathological features showing the presenceof giant cells, mononuclear infiltrates or granulomas inassociation with different features of necrotizingarteritis, and rarely fibrinoid necrosis[3, 9]. Other recognizedpathological findings are those of healed arteritis, and aredistinguished from changes seen in arteriosclerosis by thepresence of focal mononuclear aggregates and longbreaks in elastic lamella[9]. Since arterial wall inflammationis segmental, the two temporal arteries may also be unevenlyinvolved. Therefore, histological signs of inflammation maybe missed in temporal artery biopsy (TAB) performed inarteritis-free segments. As a result, in most studies 10%–20%of TABs are reported as negative in patients with GCA,although the rate may be as high as 40%[8,10]. The study by Breuer suggest that performing bilateral temporal arterybiopsies increases the diagnostic sensitivity of the procedureby up to 12.7% compared to unilateral biopsies[11], which waslikewise observed in a study by Khalifa where the rateof biopsy positive GCA increased to 73% from 64.6% whena contralateral biopsy was performed[12].Our patient's unilateral TAB of 2cm length showed intimalfibrosis of the tunica media devoid of any inflammatoryinfiltrate. This finding was also seen in 27% of cases in aseries of histopathologic findings of temporal arteritis[12].A casereport by Petzold also described a patient presentingwith 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 while some can remain positive for up to4–6 weeks after commencing high dose corticosteroid[13]. Our patient received high dose corticosteroid for one week 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 authorssuggest pursuing imaging studies such as ultrasound, CT or MR angiography of the aorta and great vessels[14].We performed complete imaging studies in our case and found high grade rightinternal carotid artery occlusion. Patients dying during the active phase of GCA have a high incidence of involvement of the superficial temporal, vertebral, ophthalmic, and posterior ciliary arteries, whereas the internal carotid, external carotid, and central retinal arteries are less frequently involved. When the carotid artery is involved, the siphon is the most frequently involved segment[15].Since the distal part of the interacranial oculomotor nerve receives nutrient arterioles from inferior cavernous sinus artery , third nerve palsy can be caused due to involvement of this branch of internal carotid artery .

CONCLUSION

Diagnosis of temporal arteritis is often delayed due topoor recognition of early, often non-specific symptoms.Hence, it should be considered in all patients (even in those with ESR <50and negative biopsy) presenting with temporal artery related symptomsor signs.GCA can present with acute third nerve palsy, mimicking the presentation of a microvascular cause. Early diagnosis is crucial becauseit can potentially cause devastating neuroophthalmiccomplications once treatment is delayed.

Conflict of interest: The authors declare no conflict of interest.

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