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Giant Cell (Temporal) Arteritis with Lingual Necrosis ： A Case Report

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Abstract

Giant cell (temporal) arteritis (GCA) is a granulomatous inflammatory disease affecting particularly large and medium sized arteries. Most cases occurred in the Scandinavian elderly greater than 50-year-old. The American Africans and the oriental have a much lower incidence of GCA. Headache, fever and anemia are common symptoms. Blindness is the most dreaded complication. Reported cases of definite GCA with biopsy among local population are rare. Its association with lingual necrosis as a complication is even less well known. We describe a case of biopsy proven GCA in a 93-year-old man who presented with masticatory claudication and severe temporal headache. Lingual necrosis resulting in partial glossectomy occurred despite of prompt improvement of headache after starting treatment with 40 mg of prednisolone on the second day. (J Intern Med Taiwan 2006; 17: 118-122)

Key Words ： Giant cell arteritis, Temporal arteritis, Lingual necrosis

Introduction

Giant cell (temporal) arteritis is a granulomatous arteritis the cause of which is not well delineated. It is thought to be immunologically mediated but no consistent or significant immunologic abnormalities have been reported [1]. Of all the complications of GCA, the loss of eyesight is probably the most serious [2]. Lingual necrosis has been only reported rarely in the literature as a complication of GCA. For some reason GCA occurs in the white population almost exclusively[3]. Most reports on GCA originate from Scandinavia and from parts of the northern United States in populations of the same ethnic background[4-

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[5]. The incidence of temporal arteritis in Orientals is virtually unknown. Up to now, review of both English and Chinese literatures shows only few cases of patients of Chinese heritage with histologically proven GCA[6-9]. The accompaniment of the rare complication of lingual necrosis while the patient was receiving high dosage of steroid treatment was even rarer or not reported in this population.

Case Report

A 93-year-old man was admitted to the hospital for severe headache. About one month prior to admission, patient developed a left temporal headache. The headache was low grade, intermittent and throbbing in nature. About three days prior to admission, patient developed intermittent masticatory claudication. On the morning of admission, there was rapid aggravation of the headache and it was so bad that the patient came to the emergency service to seek for medical attention.

When seen in the emergency room, the patient was conscious, oriented, cooperative but in agony. He used his hand to cover his left peri-auricular and forehead regions. He denied any blurred vision. When palpated, the left temporal artery was tender and a bit hard in consistency to touch with no pulsation noted. Other aspects of bedside neurological examination including fundoscopy and confrontation test for visual field were normal. There was no mention of any tongue problem at that time. On admission, erythrocyte sedimentation rate (ESR) was 87 mm per hour, white blood cell (WBC) count was 11.05 x 10^9/L, and hemoglobin was 11.1 g/dl. On the second hospital day, the patient's left face was painful and was cooler to touch than the right side. The pain was so intense that the patient became very agitated and was unwilling to talk. He was started on Prednisolone (40 mg per day). Headache and facial pain improved a lot on the third hospital day and temporal artery biopsy was performed on the fourth day. On the sixth hospital day, despite a developing mild pain at the tip of left side of the tongue and a small region of grayish patch at that region, the patient requested for discharge against medical advice. He was given both oral prednisolone and topical steroid for home medication.

Patient returned on the eighth day for severe pain in the tongue. When examined, the tongue was cyanotic especially in the left side. The previous small patch of grayish coating became much larger and was necrotic and bluish-grey in color. The left face was much cooler to touch and with severe local tenderness. There was no erythematous change. Visual field examination again was grossly intact. There was no other significant focal neurological deficit. Repeated ESR on the eighth day was 45 mm per hour. The patient was started with a bolus dose of 250 mg of

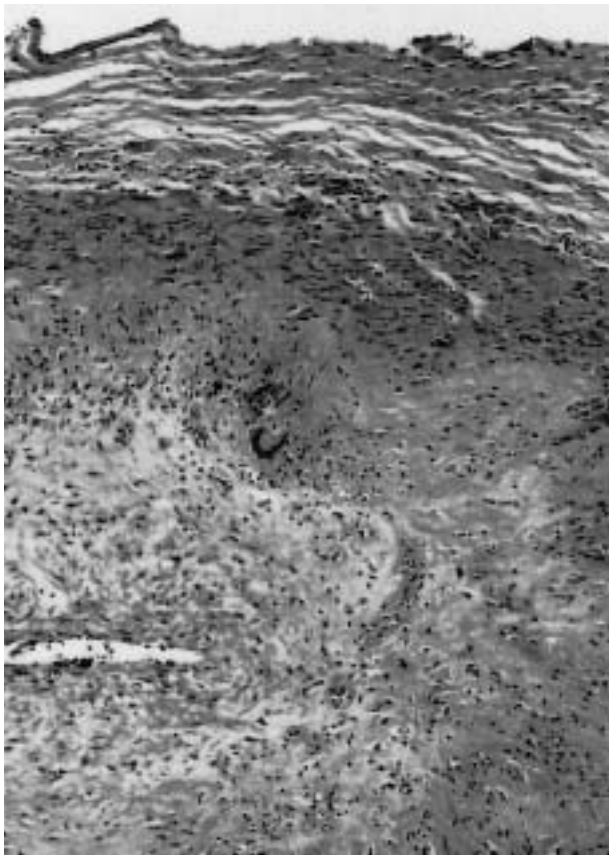

Fig.1. The temporal artery showed whole layer inflammation, especially inner media with presence of granulomas and giant cells. The intima was markedly edematous and fibrotic, resulting in extensive luminal obliteration. (Hematoxylin & eosin stain, original magnification 100 X)

methylprednisolone intravenously. Carotid artery angiography on the twelfth day demonstrated a common orifice of the innomate artery and left common carotid artery. There was diffuse narrowing of the left external carotid artery. Only the occipital and superior thyroid branches were found. The whole carotid system in the right side was patent. Due to persisting high fever and marked leucocytosis despite of strong antibiotic coverage, partial glossectomy was performed. The fever promptedly subsided one day after the operation. Histologic examination revealed gangrenous necrosis of tongue, active inflammation and luminal narrowing of nearby medium-sized arteries (Fig.1). He was discharged home on the twenty-two hospital day with home medication of 60 mg/day of oral prednisolone.

Discussion

To most physicians in their training days, they must have been taught not to miss GCA as a potential cause of headache in any elderly who presented with a recently onset unilateral headache. Any misdiagnosis and delay of treatment could result in grave consequence of rapid loss of vision in one or both eyes. However, in spite of its apparent popularity in textbooks, GCA is predominately a disease of the Caucasians. A review of both the Western and Chinese literatures showed only few cases of histologically proven GCA in patients of Chinese heritage. There have been reports of high prevalence of HLA-DR4 antigen among patients with GCA[10]. It has also been reported that the paucity of GCA in the American Africans is linked to the low frequency of HLA-DR4 in the American Africans[11]. The cause of the rarity of this disease among patients of Chinese heritage is not known. Whether the frequency of HLA-DR4 in the Orientals and Chinese might have any role to play in relation to the rare occurrence of GCA in this population remains to be clarified.

To diagnosis GCA, three out of five of the criteria set by the American college of rheumatology in 1990 must be fulfilled. These included age greater than 50; new onset of headache; temporal artery abnormal; elevated ESR and abnormal biopsy[12]. Our reported case satisfied all five of these criteria.

It has long been proposed that a cell-mediated immune response directed toward antigens in or near the elastic tissue component of the arterial wall or against the smooth muscle cell may account for pathogenesis of this disorder[13-14]. More recent reviews[15-20] confirmed that GCA is a T-cell-dependent disease[15] and that CD4+ T cells orchestrate the injury of tissues in the vasculitis process[16]. The activation of T-cells in the nonlymphoid environment of the arterial walls requires the activation of dendritic cells (the specialized antigen-presenting cells)[17]. Monocytes and macrophages are also activated and are responsible for the systemic inflammatory syndrome in GCA[18]. Depending on the unique response of the resident cells in the arterial wall to the immune injury mediated by tissue-infiltrating cells[19], occlusive vasculopathy caused by the rapid proliferation of the intima or formation of aortic aneurysm caused by the destruction of the arterial wall may result[20].

Common clinical symptoms of GCA included headache (90%), ocular symptoms (50%) and masticatory symptoms (50%)[1]. Masticatory claudication frequently is associated with bilateral facial artery occlusion. The cause of pain may be due to ischemia of the temporalis or masseter muscles. It may also be due to local pressuring effect by the mandibular condyles on the adjacent superficial temporal artery[2]. Clinical manifestations of lingual arteritis is reported to arise in 25% of patients with GCA but only few developed to lingual infarction[2]. This may be due to the rich vascular supply of the tongue. The oral symptoms and signs may include tongue swelling, recurrent blanching, recurrent pain, difficulty in tongue movement and less commonly, tongue necrosis[12]. Intermittent lingual claudication and paroxysmal blanching have been reported to precede frank necrosis and are important symptom and sign to watch out[21].

In the treatment of GCA patient, it is very important to start steroid therapy as soon as possible, sometimes even before biopsy. It is of interest that our patient developed lingual necrosis in spite of his continuous prednisolone treatment and showing improvement in other clinical parameters. The initially lower dose of prednisolone used in this case might contribute to his lingual complication. Should a higher dose of possibly 1 mg/kg be used, the complication of lingual infarction might be able to be averted.

This case also serves to remind of the importance of watching out for the development of further complications even if the headache appears to be improving. It has also been reported that some drugs including ergotamine for treatment of migraine[22] and cold-induced vasospasm[2] would precipitate the development of lingual infarction. However, our patient did not take any ergotamine like medications and it was almost summer time when the acute attack occurred.

In summary, although GCA frequents itself in the list of differential diagnosis of acute headache in the elderly, in actual daily practice, it is rarely recognized and probably underdiagnosed. We reported a case of GCA with the rare complication of lingual necrosis despite of clinical improvement of headache with lowering of ESR under steroid therapy. Early detection as well as adequately high dose of steroid might be able to save such future patient from complications.

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巨細胞動脈炎合併舌壞死：一病例報告

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摘　要

巨細胞動脈炎是一種好發於大血管及中血管的疾病，常見於『斯堪的那維亞』裔五十歲以上的患者，但在非裔美國人及東方人則相當少見。 患者常以頭痛，發燒，貧血等來表現。 嚴重者可併發有失明的併發症。本土報告有病理切片的案例並不多，而以舌頭壞死來表現者更爲罕見。我們報告一例有病理切片證實的93歲男性病人，在發病後雖以類固醇治療頭痛有改善，但最後仍有舌頭壞死的併發症，不得已需以外科手術切除。