

CASE REPORT

Giant Cell Arteritis Manifested By Bilateral Arteritic Anterior Ischemic Optic Neuropathy and Jaw Pain: A Visually Devastating Condition*Mrunal Kurangi^{1*}, Ugam Usgaonkar¹**¹Department of Ophthalmology, Goa Medical College, Bambolim- 403202 (Goa) India***Abstract**

Giant Cell Arteritis (GCA) is a systemic vasculitis of unknown etiology affecting medium and large calibre vessels by granulomatous panarteritis with the formation of giant multinucleate cell granulomas. Vision is affected in 25-50% of GCA patients. Affection of vision may be the first GCA symptom or a symptom which occurs weeks or months after the initial symptoms of the disease. Permanent damage to the patient's vision is a serious consequence of GCA. Arteritic Anterior Ischaemic Optic Neuropathy (AION) is the most frequent and most serious visual manifestation of GCA. It is manifested by partial or total loss of vision. Arteritic AION therapy in GCA uses high doses of glucocorticoids, but glucocorticoid therapy has a number of adverse effects. The proofs of the effect of the therapy on the improvement of the vision of patients with visual affection in GCA are not convincing. We report a case of a 55-year old female with biopsy-verified GCA whose primary manifestation was bilateral arteritic AION resulting in a complete loss of vision in one eye and dramatic worsening of visual acuity in the other eye. Even after diagnosis glucocorticoid therapy could not salvage her vision and led to secondary complications. This proves that appropriate high dose glucocorticoid therapy does not promise success of therapy in GCA. An increased awareness of giant cell arteritis should lead to earlier diagnosis and treatment and avoidance of the devastating consequences.

Keywords: Arteritic Anterior Ischemic optic

Neuropathy, Vasculitis, Giant Cell Arteritis, Glucocorticoids

Introduction:

Giant Cell Arteritis (GCA) is the most common primary vasculitis of adults in the Western world [1]. The worldwide incidence of this condition ranges from 1 to 29 per 100 000 persons 50 years of age or older, which is similar to the age-adjusted incidence rates for oral cancers [2]. GCA occurs almost exclusively in individuals older than 50 years of age and is more common among women than men (ratio of 3:1) and among those of Scandinavian ethnicity [3]. The clinical manifestations are diverse, and patients may present to various specialists, including rheumatologists, neurologists, cardiologists, ophthalmologists and dentists. Typical clinical features may include fever, headache, jaw claudication, palpable and tender temporal arteries, symptoms of Polymyalgia Rheumatica (PMR), visual disturbance, and raised Serologic inflammatory markers such as the erythrocyte Sedimentation Rate (ESR) or C Reactive Protein (CRP). The extracranial and intracranial branches of the carotid artery are frequently affected, which accounts for the craniofacial, oral and ocular symptoms of the condition. Moreover, serious and life-threatening presentations, such as permanent

loss of vision and cerebrovascular stroke, are relatively common [4-6].

GCA can be an elusive diagnosis because of the variable presentation. However, the potential for devastating consequences should prompt care providers to consider it as a possible diagnosis. This article describes a woman whose primary manifestation was bilateral arteritic AION resulting in a complete loss of vision in one eye and dramatic worsening of visual acuity in the other eye along with jaw pain. Even after diagnosis glucocorticoid therapy could not salvage her vision and led to secondary complications. This proves that appropriate high dose glucocorticoid therapy does not promise success of therapy in GCA. An increased awareness of giant cell arteritis should lead to earlier diagnosis and treatment and avoidance of the devastating consequences.

Case Report :

A 55-year-old woman presented to the Open Patient Department with a sudden painless diminution of vision in her left eye since 10 days. On direct questioning, she denied headache or scalp tenderness. She had significant jaw pain that had limited her diet to soft, pureed foods. No clear cause of the jaw pain had been identified before. Moreover, systemically she was earlier diagnosed as a case of diabetes mellitus type II, systemic hypertension and chronic kidney disease since last 2 years for which she was on appropriate medications and dialysis. During the current episode, she was seen by the ophthalmologist. She had only light perception in the left eye and 20/20

acuity in the right eye. There was an afferent papillary defect on the left. The visual fields could not be checked on the left, but visual fields by Humphrey's analyser were mildly constricted with enlargement of blind spot on the right (Fig. 1). Examination of the fundus revealed elevated, blurred disc margins with vessel tortuosity and disc hemorrhages in both eyes (Fig. 3 and Fig.4). Both fundi also showed signs of moderate non proliferative diabetic retinopathy. Patient was admitted as bilateral Arteritic Anterior Ischaemic Optic Neuropathy (AION) under investigation. Her systemic examination did not reveal temporal artery abnormality.

The following day, visual acuity had declined dramatically in both eyes. The patient was unable to perceive light in her left eye and could read 20/200 only with the right eye. The repeat visual fields of the right eye showed a centroca ecalscotoma (Fig. 2). Inflammatory markers were measured and both ESR (63 mm/h) and CRP (24 mg/L) were elevated. At this facility, the normal range for erythrocyte sedimentation rate was 5–20 mm/h, but the upper limit of normal is age-dependent and can be approximated as age divided by 2 (for men) or (age + 10) divided by 2 (for women). CRP is typically undetectable in the blood, and any value above 3 mg/L is considered elevated. The patient was labelled as GCA based on clinical findings. High-dose intravenous methylprednisolone therapy was initiated (250 mg 4 times a day).

Biopsy of the temporal artery confirmed the diagnosis of GCA (Fig.5). The patient's vision

continued to decline in the left eye, despite steroid therapy. After the 3-day course of methyl prednisolone, she was unable to perceive light in either eye, although her jaw pain had diminished dramatically. She was switched to a maintenance dose of oral prednisone, which was to be slowly tapered. But unfortunately she developed respiratory tract infection and uncontrolled blood sugar levels for which she required Medicine expert opinion for systemic antibiotics, insulin therapy and routine dialysis.

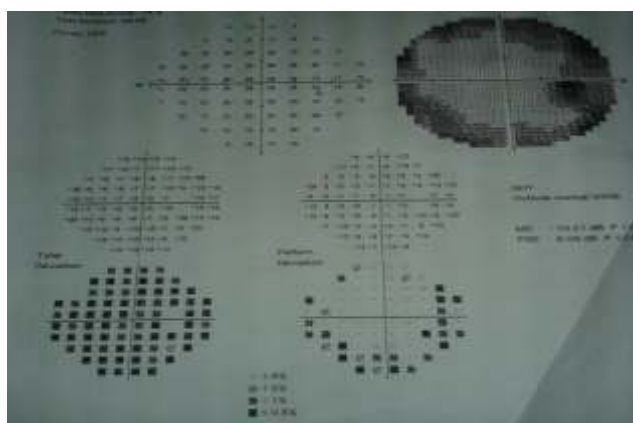


Fig. 1 : Right Eye Fields on HFA showing Mildly Constricted Fields with Enlargement of Blind Spot

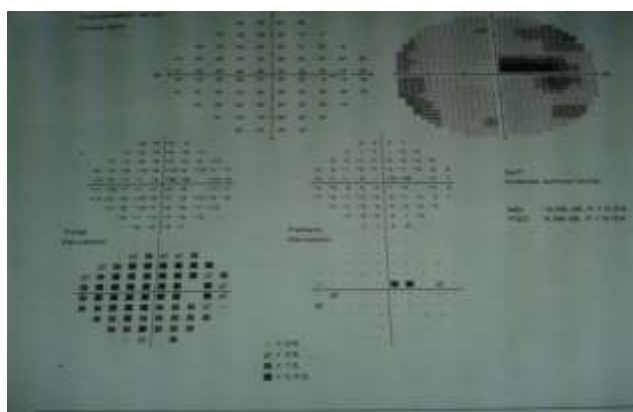


Fig. 2 : Repeat Fields of Right Eye showing a Centrocaecalscotoma

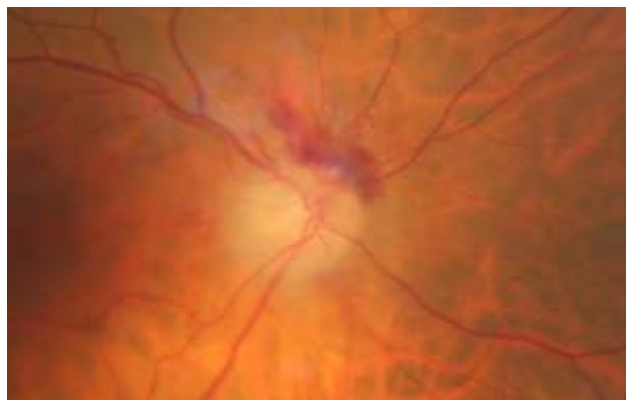


Fig. 3: Right Eye Fundus Picture showing Pallid Edematous Disc with Hemorrhages

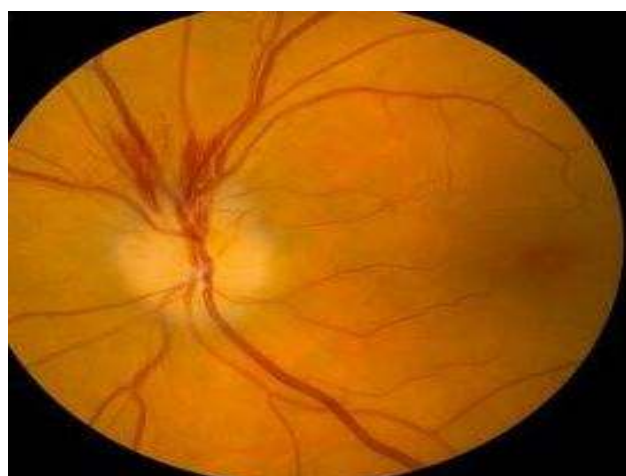


Fig. 4 : Left Eye Fundus Picture showing Similar Pallid Disc Edema

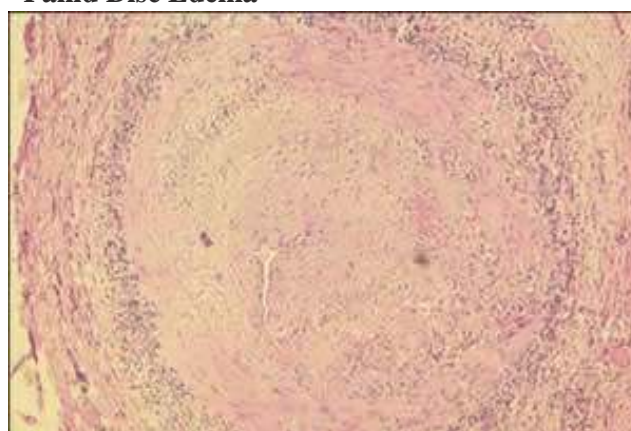


Fig. 5 : Histopathological Section of Temporal Artery Biopsy showing Intense Inflammatory Changes in Vessel Wall Media with Luminal Constriction

Discussion:

The diagnosis of GCA is based on clinical criteria outlined by the American College of Rheumatology [7] (Table 1). The presence of 3 of these 5 features permits a diagnosis of GCA with a sensitivity of 93.5% and specificity of 91.2% [8]. In the case reported here, the patient's age at onset, the elevated erythrocyte sedimentation rate and the abnormal results of temporal artery biopsy confirmed the diagnosis. On presentation, she denied any headache and did not have any appreciable abnormality of the temporal artery on palpation, but she did have a dramatic change in her vision.

Claire A. Sheldon performed a literature review of articles published from January 1, 2005, to January 1, 2010, and recorded the presenting symptoms in 2011. In 81 studies, involving a total of 390 patients, visual symptoms were reported in 28.5% of cases (Table 2) [18]. In patients older than 50 years of age, GCA should be considered in the differential diagnosis of a variety of ocular symptoms, such as transient visual blurring, diplopia, monocular visual field loss and profound vision loss [6]. The vision loss associated with GCA is often irreversible, and treatment with high-dose steroids is aimed at both minimizing further declines in vision and preventing contralateral vision loss [9, 10]. In a large retrospective analysis, 14% of patients with biopsy-confirmed GCA had experienced permanent vision loss [11]. Among patients

whose GCA was left untreated, 25%–50% experienced contralateral loss of vision over the following 1–2 weeks [11]. However, after initiation of steroid treatment, the risk of progressive vision loss was just 1% among patients who had not had visual symptoms and 13% among those with previous vision loss [11]. These observations underscore the point that early diagnosis and treatment of GCA can prevent permanent loss of vision.

In table 1, diagnostic criterion for giant cell arteritis, as defined by the American College of Rheumatology [7]

- Age at onset of disease > 50 yr
- Localized headache (of new onset or of a new type)
- Abnormality of the temporal artery, specifically tenderness to palpation or decreased pulsation, unrelated to arteriosclerosis of cervical arteries
- Erythrocyte sedimentation rate > 50 mm/h
- Abnormal results on biopsy of temporal artery : artery showing vasculitis characterized by a predominance of monocular cell infiltration or granulomatous inflammation, usually with multinucleated giant cells

The presence of 3 of these 5 features permits a diagnosis of giant cell arteritis with high sensitivity and specificity.

In the case reported here, the patient's loss of vision was preceded by symptoms consistent with the diagnosis of GCA. First, she had a history of jaw pain severe enough to motivate a change in diet. Although a variety of symptoms of GCA may prompt presentation to a dentist's office, including jaw pain, trismus develops after a few minutes of chewing.^[8] This diff, dysphagia, glossodynia and tongue ulceration [11], jaw pain was reported in 31.3% of cases previously noted literature review, summarized above (Table 1)[18].

Table 1: Frequency of Symptoms of Giant Cell Arteritis in 390 Cases Reported in 81 Studies

Presenting symptom	No. (%) of cases
Headache	230 (59.0)
Jaw pain or claudication	122 (31.3)
Visual symptoms	111 (28.5)
Scalp pain or necrosis	89 (22.8)
Oral symptoms	28 (7.2)
Systemic symptoms	173 (44.4)

It is necessary to distinguish the jaw claudication associated with GCA from TMJ pain (Table 2). For example, jaw pain associated with TMJ begins immediately upon movement of the jaw, but jaw pain associated with GCA erence reflects a difference in the mechanism of pain: TMJ jaw pain relates to a mechanical problem, whereas GCA develops secondary to vascular occlusion and ischemia of the masseter muscle. Moreover, GCA typically occurs in patients over 50 years of age, but TMJ jaw pain is most common in patients

between 25 and 44 years of age and occurs in the absence of an associated headache and constitutional symptoms [12].

An accurate assessment of GCA-associated jaw pain can lead to a timely diagnosis. Jaw claudication is the symptom of GCA most often associated with a positive result on temporal artery biopsy, which indicates that the presence of jaw claudication has a high level of diagnostic sensitivity [8]. The presence of jaw pain has also been linked to an increased risk of permanent loss of vision [9]. Similarly, the presence of trismus, a rare manifestation of GCA, may indicate more aggressive disease [13].

This case highlights the importance of considering the diagnosis of GCA. Additional cases of jaw pain related to GCA being misdiagnosed as TMJ pain have been reported in the literature [12, 15]. However, in contrast to the case presented here, those patients did not experience permanent loss of vision. If left untreated, GCA can have devastating consequences. Early diagnosis is essential to avoid these preventable conditions [4-6, 16, 17].

Table 2: Comparison of Clinical Features of Jaw Pain Associated with Temporomandibular Joint Pathology and Giant Cell Arteritis

Temporomandibular Joint Pathology	Giant Cell Arteritis
Unilateral or bilateral dull aching pain localized to the area of the jaw, neck, ear and temple	Jaw claudication
Immediate onset of pain with movement of jaw	Onset of jaw pain after a few minutes of mastication; pain disappears with rest
Age 25–44 yr	Age > 50 yr
Associated tenderness and spasm of muscles of mastication and possible limitation of jaw movement	May be associated with swelling of the ipsilateral face, tongue and buccal mucosa and (rarely) necrosis of the tongue
Response to heat, massage, salicylates, muscle relaxants and occlusal splints	Additional features: headache, scalp tenderness and systemic symptoms (e.g., fever, anorexia, weight loss)

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