

# Giant cell arteritis or tension-type headache?: A differential diagnostic dilemma

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## ABSTRACT

Giant cell arteritis (GCA) or Temporal arteritis (TA) is an autoimmune disease and the most common type of vasculitis in the elderly. It causes inflammation of the medium and large arteries in the upper part of the body. GCA is an under-recognized cause of head aches in the elderly, especially when it presents itself with atypical features, resulting in delayed or incorrect diagnosis. Since GCA is a treatable condition, an accurate diagnosis is crucial to prevent the most serious complication of CGA, permanent vision loss. The diagnosis can be further complicated as GCA may present with features of other painful neurological conditions. The present case is an 81-year-old woman diagnosed with GCA, who initially presented with features similar to tension-type headache. Due to overlapping features of these conditions, the diagnosis of GCA was delayed, resulting in irreversible vision loss. Although previous research highlights diagnostic dilemmas featuring GCA and other disease states, this case is exclusive in describing a unique dilemma where tension-type headache mimics GCA.

**Key words:** Giant cell arteritis, headaches, temporal, tension-type headache, vasculitis, vision loss

## Introduction

Giant cell arteritis (GCA), also known as temporal arteritis (TA), is an autoimmune disease and the most common form of vasculitis in individuals over the age of 50.<sup>[1]</sup> It is regarded as one of the possible underlying causes of secondary headaches in the elderly.<sup>[2]</sup> In terms of harmful effects, it is considered a neurological emergency and it is recommended that upon clinical suspicion, treatment be initiated promptly to prevent irreversible damage.<sup>[3]</sup> GCA involves medium and large arteries in the head, neck, and upper part of the body. Branches of the external and internal carotid arteries are particularly susceptible and their involvement leads to the classic manifestations of visual loss, headache, scalp tenderness, and jaw claudication.<sup>[1]</sup> Jaw claudication is a prominent symptom of GCA described as pain in the jaw

during mastication or speaking and results from ischemic maxillary artery supplying the masseter muscles. The headache is described by patients as throbbing in nature and usually occurs in both temples. The prevalence is higher in Scandinavian countries, which is twice as much as other European countries. The age of onset of GCA is usually greater than 50 years; however, it is most commonly seen in patients between 70 and 80 years of age.<sup>[1]</sup> Females have a higher incidence than males with a female to male ratio of 3:1.<sup>[2]</sup>

Tension-type headache is classified by the International Headache Society as a primary headache. This pain originates typically in the forehead and bilaterally spreads to the occiput, ultimately reaching the neck muscles.<sup>[4]</sup> The pain is classified under the category of tightening and non-pulsating and is not accompanied by nausea or any other elements of migraine symptomology.<sup>[4]</sup> Due to its nature as a primary type, tension-type headaches are not caused by any endogenous neural dysfunction or disease, whereas GCA is caused by inflammation of the arteries. A detailed history and physical examination is essential for accurate diagnosis, especially if GCA presents with atypical features. Since GCA is a treatable condition, it is important to make the correct diagnosis

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to prevent the potential complication of irreversible vision loss. Corticosteroids remain the gold-standard treatment for GCA.<sup>[1]</sup>

Herein, we report a case of an elderly female, who presented with clinical features akin to tension-type headache one year prior to her correct diagnosis of GCA.

## Case Report

An 81-year-old woman was referred to our clinic with the chief complaint of a newly developed onset headache, which occurred on a daily basis and was localized in the left occipital area. The pain would radiate toward her lower mandible area and was stabbing in nature. However, a few months later, the pain involved the parietal region as well. The pain was nocturnal, causing her to wake up 3 to 4 times on average per night. Furthermore, the patient described the headaches as throbbing in nature, no nausea or vomiting, no history of light sensitivity, and no aura or focal motor or sensory symptoms. She also had no trouble with vision and no tenderness of the temporal arteries. Her neurological examination was unremarkable. A CT scan of the head displayed moderate cerebral atrophy but was otherwise unremarkable. She continued to use over-the-counter analgesic medications with temporary relief of her headaches. One year later, she was presented to the ophthalmology clinic with left visual disturbances. Her left and right visual acuity was 20/300 and 20/30, respectively. In addition to this, she had symptoms of bilateral jaw claudication and an elevated CRP of 59. She subsequently underwent a temporal artery biopsy on the left side, which was positive for GCA. She was treated with corticosteroids and responded well to the treatment; however, her visual acuity did not improve.

## Discussion

The exact etiology of GCA is unknown. The inflammation of large- and medium-sized arteries in GCA is thought to be due to an antigen-mediated autoimmune response.<sup>[1,2]</sup>

Clinically, GCA has a broad spectrum of associated symptoms, with the most common being headache (70-90%), jaw claudication (40-60%), transient ischemic attack (4%), neck pain, scalp tenderness, and visual disturbances.<sup>[1]</sup> The American College of Rheumatology (ACR) has devised a set of criteria for temporal arteritis which requires that at least three of the following conditions be met: (i) patient age >50 years, (ii) new onset of headache, (iii) temporal artery tenderness, (iv) elevated ESR, and (v) abnormal

findings of temporal artery biopsy.<sup>[5]</sup> However, ACR criteria are utilized best for classification purposes and are not validated for use in clinical diagnosis. Interestingly, our patient had only occipital pain without developing any temporal pain. This is an atypical presentation of GCA, which potentially contributed to the delayed diagnosis and her subsequent serious complication of irreversible visual loss.

The visual symptoms of GCA can include amaurosis, diplopia, and visual loss which are due to ischemic optic neuropathy, and visual symptoms may develop months after the initial symptoms onset.<sup>[2]</sup> About 20 to 50% of patients develop ischemia of the optic nerve, leading to irreversible blindness.<sup>[3]</sup> Before other manifestations of GCA are experienced, patients may complain of pain in their teeth, jaw, ear, or zygoma. Treatment with corticosteroids should be initiated immediately if the diagnosis of GCA is suspected. Corticosteroids are administered and improvements in systemic symptoms and normalization of CRP or ESR are noted. This change is usually seen in the first 3 to 4 weeks.<sup>[3]</sup> The duration of corticosteroid therapy is variable and can extend to several years depending upon patient needs. Our patient responded well to corticosteroids but her visual acuity did not improve. EULAR guidelines provide several recommendations and mention other pharmacological therapies that may be helpful in the treatment of GCA.<sup>[6]</sup> Furthermore, there is evidence showing a rapid response with the use of tocilizumab, an IL-6R antagonizing monoclonal antibody, including normalization of GCA symptoms and inflammatory markers.<sup>[7]</sup> Despite these encouraging results, randomized control trials are underway to fully confirm its efficacy in the treatment of GCA.

GCA as a secondary headache has been subject to misdiagnoses when presented with atypical features that are more representative of other disease states as seen in our case. This has been supplemented through further evidence of other GCA diagnostic dilemmas in the literature. One case report presented an elderly patient suffering from orofacial pain, which is an atypical feature of GCA.<sup>[8]</sup> Subsequently, the patient was misdiagnosed with temporomandibular disorder.<sup>[8]</sup> In addition, Kraemer *et al.* emphasized the importance of greater analysis into individual GCA symptoms.<sup>[9]</sup> This was elucidated in their case report as the reduction of jaw opening (i.e. an atypical feature) in their patient led to the misdiagnosis of GCA as occipital neuritis and craniomandibular dysfunction.<sup>[9]</sup> Furthermore, GCA can also be associated with other diseases such as Takayasu arteritis.<sup>[10]</sup> Despite the literature being laden with several diagnostic dilemmas concerning GCA, our case report is

unique in terms of being the only one where GCA was misdiagnosed as tension-type headache.

It has been noted that tension-type headache can present itself as GCA.<sup>[11]</sup> However, our case presents an opposite dilemma, as our patient presented with atypical features of GCA that were more characteristic of tension-type headache. That is, GCA presented itself as tension-type headache. Hence, it is important to be aware of overlapping features of these conditions to prevent misdiagnoses. Furthermore, a lack of clinical findings concerning GCA in general can lead to an increase in misdiagnosed cases. Most importantly, there is a lack of robust findings in literature concerning cases where GCA presents itself as tension-type headache, which underscores the distinctiveness of our case.

Lastly, our case emphasizes the importance of considering GCA as a potential diagnosis for elderly patients with new onset headache located in a non-temporal region. Our patient had only occipital/parietal pain throughout the course of the disease, without the involvement of any temporal pain. In order to resolve similar future diagnostic dilemmas, we recommend these patients should have a full workup for GCA conducted as well as checking their ESR and CRP levels. They need to be screened immediately upon clinical suspicion of the disease. This may prevent the severe complications of irreversible vision loss, as was seen in our case. Although the diagnosis of GCA can be impeded due to the conflicts of its clinical presentation with several other neurological

conditions, this unique case highlights the importance of considering GCA and its atypical characteristics when patients present with features similar to tension-type headache.

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