Giant Cell (Temporal) Arteritis with Persistent Bilateral Sensorineural Hearing Loss – A Likely Consequence of Delayed Institution of Glucocorticoid Therapy

ACDEF 1 Shoaib Junejo
ACDEF 2 Yasir Ali
AF 1 Adriana Abrudescu

Corresponding Author: Shoaib Junejo, e-mail: shoaibjunejo88@gmail.com

Conflict of interest: None declared

Patient: Male, 66
Final Diagnosis: Giant cell arteritis
Symptoms: Hearing loss
Medication: —
Clinical Procedure: —
Specialty: Rheumatology

Objective: Unusual clinical course
Background: The classical picture of giant cell (temporal) arteritis (GCA) is not always evident. A wide variety of unusual presentations have been increasingly reported. Sensorineural hearing loss (SNHL) as an initial manifestation of GCA is a rare, yet important entity. Similar to blindness, SNHL also deserves serious attention as timely intervention may play a key role in overall prognosis. Only a few cases of SNHL associated with GCA have been reported in the literature, the majority of them were diagnosed early and responded well to glucocorticoid therapy. Our report focuses on a case of persistent audiometry-proven SNHL despite optimal doses and duration of glucocorticoids required in GCA patients with ischemic complications.

Case Report: We present the case of a 66-year-old male with severe bi-temporal headache, left jaw claudication, and progressively worsening hearing loss, the latter symptom reported over the preceding year. Examination of his temporal artery remained insignificant, but the laboratory data showed raised erythrocyte sedimentation rate (ESR) of 52 mm/hour. Audiometry performed and the interpretations were coincided with the bilateral high frequency mild to moderately-severe SNHL. Prompt administration of IV methylprednisolone started at high-optimum doses that were gradually tapered over the subsequent six months. On biopsy of the left temporal artery, the findings were consistent with the GCA. The patient responded well in terms of headache and general condition improvement, but results of repeat audiometry at follow-up visits proved disappointing and the interpretations were that there was no change in the audiometry results based on first presentation.

Conclusions: This case is purposefully reported to draw the attention of practicing physicians, and encourage them not only to better understand atypical presentations of GCA but also to intervene in a timely fashion. This case should encourage literature to reset recommendations and encourage have high-indexed suspicion when elderly patients present with deafness since early diagnosis and treatment may have profound effects on overall long-term prognosis of other cranial ischemic complications as well.

MeSH Keywords: Audiometry • Hearing Loss, Bilateral • Hearing Loss, Sensorineural • Temporal Arteries

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/907174
Giant cell (temporal) arteritis (GCA) is a chronic systemic vasculitis affecting typically large- and medium-sized vessels [1]. The disease is multifactorial with aging posing the greatest risk. Essentially, it rarely occurs in individuals before the age of 50, the incidence steadily rises thereafter, with the peak achieved in the seventh decade of life [2,3]. Epidemiologically, GCA is the most common form of vasculitis across the Northern hemisphere latitude; the annual incidence reported highest in Scandinavia and North America with the rate greater than 17 cases per 100,000 people per population age ≥ 50 versus the rate of lower than 12 cases per 100,000 people per population of the Southern Europe and Mediterranean countries with the same age group [3–5]. Since GCA has a considerable predilection to involve cranial branches evolving from the aortic arch [6], the classical presentation is an elderly patient with new onset of headache, scalp tenderness, jaw claudication, and blindness. Headache is the most common symptom whereas, 40% to 50% of patients have associated polymyalgia rheumatica (PMR) [7]. Patients with other atypical presentations have been reported, but only a handful of cases manifest audiovestibular symptoms have been reported until now. Our case report is supported by a 10-year retrospective study where 271 biopsy-proven GCA cases were taken into the account [8]. The facts displayed the retrospective study were: 1) the rarity of sensorineural symptoms in GCA patients, only 1.5% (4 of 271 cases), 2) the symptoms may either precede – solely or appear concomitantly – with other features of GCA, and finally 3) the desired response to glucocorticoid therapy is likely time-dependent. All four cases, in the aforementioned study had biopsy proven diagnosis of GCA within a three-month period from the onset of hearing loss and responded well to treatment. Based on this finding, we made the assumption that treatment failure in our patient case could be related to the delayed diagnosis and institution of glucocorticoids since he took a year to seeking the medical attention.

Case Report

A 66-year-old male patient presented with a four-week history of worsening bi-temporal headache and progressive hearing loss over the preceding one-year. He remained afebrile and denied symptoms, such as vision changes, photophobia, phonophobia, tinnitus, or weight loss but, he did report left jaw claudication typically associated with cold fluid intake, and non-specific symptoms of nasal congestion with post-nasal drip. The pain was further explained as sharp-cutting, lasting for about 10 minutes and aggravated by lying down. At presentation, his body temperature was 98.2°F (36.8°C), blood pressure 144/82 mm Hg, heart rate was regular at 71 beats/minute and respiratory rate 16 breaths/minutes with 100% O₂ saturation. On physical examination, we found the temporal and occipital arteries were normal with no evidence of overlying tenderness, nodularity or reduced or absent pulsations, and the conjunctivae appeared normal. The patient was admitted with high-indexed suspicion of GCA.

Laboratory data showed the erythrocyte sedimentation rate (ESR) 52 mm in the first hour, hemoglobin 12.6 g/dL, WBC 5.1×10³/L, C-reactive protein 4.64 mg/dL, and serum electrolytes and liver function test remained within the normal limits. Computed axial tomographic brain scan was also normal. Evaluation by an Otolaryngologist was unremarkable except for the signs of sensorineural hearing loss (SNHL). Audiometry test results were significant for the bilateral high frequency mild to moderately severe SNHL (Figure 1). We began prompt administration of IV methylprednisolone, started at 250 mg every six hours. Findings on the biopsy of the left temporal artery were consistent with the diagnosis of GCA (Figure 2). The patient was discharged on prednisone 60 mg daily, which was gradually tapered over the subsequent six months. At follow-up visits, the headache had completely subsided and ESR showed a down-trend but the patient still had hearing difficulties. The repeat audiometry showed disappointing results and the findings were similar to that of the first audiometry (Figure 3). The patient was adequately counseled about the nature of his disease and given a strong recommended to visit the emergency department in case of any dreadful symptom(s). Meanwhile, we also recommended medical follow-up with repeat ESR and annual audiological evaluations.

Discussion

The vasculitides are a heterogeneous group of disorders characterized by the cytokine-induced reactive damage and loss of vessel integrity with resultant ischemic necrosis of downstream tissues. GCA is one of its common entities which due to shared allele and sequence polymorphism of HLA-DR genes, is highly associated with PMR. Cell-mediated immune response plays a vital role in the disease pathogenesis as is indicated by the intramural trafficking of inflammatory cells, in particular CD 4+ T-cells, macrophages, and multinucleated giant cells, with enormous amount of cytokine production such as interleukin (IL)-6 and tumor necrotic factor-alpha (TNF-alpha) [9–11]. IL-6 is closely associated with the appearance and severity of disease symptoms and its significant contribution is proved by the rapid drop of plasma IL-6 levels, with the resultant symptomatic recovery with the administration of glucocorticoid therapy followed by an immediate rise of its levels once the glucocorticoids are abruptly stopped [12]. Undoubtedly, amongst cranial ischemic complications (CICs), the most threatening are stroke and permanent loss of vision, the incidence of the latter ranging from 15% to 20% [13]. SNHL is very rare, but the
equally dreadful presentation of GCA requires serious attention to be given to this complication.

Cooke was the first to report the unusual presentation of SNHL with GCA in a small series of patients discussed in the British Medical Journal [14], and currently only a few similar associations have been reported in the literature. SNHL may appear as an initial and sole manifestation of GCA disease, however, early recognition and treatment may be crucial in preventing the CICs [8,15]. The possible mechanisms of deafness in GCA patients are explained by inflammatory arthritis changes of the posterior circulation and/or terminal cochleovestibular vasculature [16]; temporal artery biopsy may appear inconclusive in such cases.

Francis and Boddie reported a case of GCA with a negative temporal artery biopsy. The patient presented with a four-week history of headache and a worsening deafness for the preceding 48-hours. The temporal artery examination and hemoglobin level appeared normal, but the ESR was significantly high. Within 24-hours of commencement of glucocorticoids, the headache disappeared and hearing returned to normal, which was then confirmed by repeat audiometry [17]. This case adds one more example that supports the need for early administration of therapy. On the contrary to aforementioned case, one large study on 240 patients emphasized that anemia and abnormal temporal artery on examination were the two best predictive factors reflecting imminent severe ischemic complications; detailed arterial examination, in another case report of GCA and associated SNHL, was found to be thickened and tender with notably reduced pulsation [7,18].

The occurrence of audiovestibular dysfunction is more frequent in GCA cases than in PMR [19]. Elderly patients and

Figure 1. First Audiometry Test Results. Right: Hearing within normal limits at 250 Hz to 3 kHz, mild to moderately severe sensorineural loss at 4 kHz to 8 kHz. Left: Hearing within normal limits at 250 Hz to 2 kHz, mild to moderately severe sensorineural loss at 3 kHz to 8 kHz.

Figure 2. Histopathological findings of left temporal artery biopsy. Trafficking of inflammatory cells is consistent with giant cell (temporal) arteritis.
patients with isolated PMR who present with new onset of audiovestibular symptoms, particularly SNHL, have a high-indexed of suspicion of underlying GCA and should be consid- ered for treatment with glucocorticoids which should be start- ed promptly [20,21]; treatment need not to be withheld while awaiting the performance or the results of the biopsy. If the temporal or other artery biopsies prove insignificant for any evidence of inflammatory vasculitis but the clinical suspicion of GCA still remains high, the glucocorticoid treatment should be continued [22].

Conclusions

This case illustrated the importance of paying serious atten- tion to SNHL in elderly patients since it may be a presenting feature of GCA. Clinicians must be updated of this possibili- ty because in addition to permanent hearing loss, early rec- ognition may also help prevent other dire complications such blindness, stroke, and aortic dissection.

Acknowledgment

Deborah Goss, MLS (Director, Health Sciences Library, Queens Hospital Center) reviewed and edited the manuscript.

References:

1. Hellmann DB, Hunder GG: Giant cell arteritis and polymyalgia rheumatica. In: Kelley’s Textbook of Rheumatology, 7th, Harris ED, Budd RC, Firestein GS, Genovese MC et al. (eds.), WB Saunders Company, Philadelphia, 2005
2. Kermani TA, Schäfer VS, Crowson CS et al: Increase in age at onset of giant cell arteritis: A population-based study. Ann Rheum Dis, 2010; 69: 780–81
3. Gonzalez-Gay MA, Vazquez-Rodriguez TR, Lopez-Diaz MJ et al: Epidemiology of giant cell arteritis and polymyalgia rheumatica. Arthritis Rheum, 2009; 61: 1454–61
4. Gonzalez-Gay MA, Miranda-Filloy JA, Lopez-Diaz MJ et al: Giant cell arteritis in northwestern Spain: A 25-year epidemiologic study. Medicine (Baltimore), 2007; 86: 61–68
5. Bas-Lando M, Breuer GS, Berkun Y et al: The incidence of giant cell arteri- tis in Jerusalem over a 25-year period: Annual and seasonal fluctuations. Clin Exp Rheumatol, 2007; 25: 515–17
6. Klein RG, Hunder GG, Stanson AW, Shpfs SG: Large artery involvement in giant cell (temporal) arteritis. Ann Intern Med, 1975; 83: 806–12
7. Gonzalez-Gay MA, Barros S, Lopez-Diaz MJ et al: Giant cell arteritis: Disease patterns of clinical presentation in a series of 240 patients. Medicine (Baltimore), 2005; 84: 269–76
8. Hausch RC, Harrington T: Temporal arteritis and sensorineural hearing loss. Semin Arthritis Rheum, 1999; 28: 206–9
9. Weyand CM, Goronzy JJ: Medium- and large-vessel vasculitis. N Engl J Med, 2003; 349: 160–69
10. Lie JT: Histopathologic specificity of systemic vasculitis. Rheum Dis Clin North Am, 1995; 21: 883–909
11. Weyand CM, Goronzy JJ: Arterial wall injury in giant cell arteritis. Arthritis Rheum, 1999; 42: 844–53
12. Roche NE, Fulbright JW, Wagner AD et al: Correlation of interleukin-6 production and disease activity in polymyalgia rheumatica and giant cell arteritis. Arthritis Rheum 1993; 36: 1286–94
13. Nesher G, Berkun Y, Mates M et al: Risk factors for cranial ischemic complications in giant cell arteritis. Medicine (Baltimore), 2004; 83: 114–22
14. Paulley JW, Hughes JP: Giant-cell arteritis, or arteritis of the aged. Br Med J, 1960; 2(5212): 1562–67
15. Imran TF, Helfgott S: Respiratory and otolaryngologic manifestations of giant cell arteritis. Clin Exp Rheumatol, 2015; 33(2 Suppl. 89): S-164–70
16. Sofferman RA: Cranial arteritis in otolaryngology. Ann Otol Rhinol Laryngol, 1980; 89(3 Pt 1): 215–19
17. Francis DA, Boddie HG: Acute hearing loss in giant cell arteritis. Postgrad Med J, 1982; 58(680): 357–58
18. Amoroso C, Vindigni MR, Almoni C: A case of temporal arteritis presenting with sensorineural hearing loss. Recent Prog Med, 2001; 92(12): 751–53
19. Amor-Dorado JC, Llorca J, Garcia-Porrúa C et al: Audiovestibular manifestations in giant cell arteritis: A prospective study. Medicine (Baltimore), 2003; 82: 13–26
20. Kramer MR, Nesher G, Sonnenblick M: Steroid-responsive hearing loss in temporal arteritis. J Laryngol Otol, 1988; 102(6): 524–25
21. Wilson H, Alderson D: Sudden sensorineural hearing loss: When is it idiopathic?. J Laryngol Otol, 2010; 124(6): 690–93
22. Hunder GG: Giant cell arteritis and polymyalgia rheumatica. In: Textbook of Rheumatology, 5th, Kelly WN, Harris ED, Ruddy S, Sledge CB (eds.), WB Saunders, Philadelphia 1996