Vestibulocochlear symptoms as the initial presentation of giant cell arteritis

Giant cell arteritis (GCA)—also known as temporal arteritis—is a systemic vasculitis that involves medium and large-caliber arteries such as the cranial arteries, great vessels, and aorta. Although the typical nonocular features of GCA are well known (e.g., new-onset headache, scalp tenderness, jaw claudication), vestibulocochlear presentations are rare and can mimic acute idiopathic sensorineural hearing loss (AISHL).[1] Although steroids are given for both AISHL and GCA, the lower dose and shorter duration of steroid treatment in AISHL can obscure and delay the diagnosis of GCA.[2] We report a case of GCA that presented with acute sensorineural hearing loss associated with eye pain and tenderness. Clinicians should be aware that AISHL is a diagnosis of exclusion, and the presence of ocular manifestations such as eye pain,should prompt consideration for GCA in the elderly.

Case report

A 74-year-old Caucasian male presented with acute sensorineural hearing loss of the left ear. Past medical history was significant for gross total resection of cutaneous squamous cell carcinoma above the left ear. He had prior mild to moderate, symmetric, age-related sensorineural hearing loss treated with hearing aids. Ocular history was significant for bilateral laser iridotomy for narrow angles. Additionally, he had a prior history of migraines, hypertension, and hyperlipidemia, which were treated medically.

The patient initially developed sudden, acute-onset, left-sided hearing loss superimposed on his prior mild to moderate, chronic, bilateral hearing loss. This was followed by left eye pain that was tender to touch. He denied headache, jaw claudication, fever, arthralgia, malaise, loss of appetite, tenderness around the temporal artery, or visual disturbances. A general ophthalmologist found a normal eye examination. Over the next few days, the patient developed worsening vestibular symptoms, which included intermittent dizziness and vertigo.

The patient subsequently was seen by an otolaryngologist who found new-onset, asymmetric, low-frequency (250 Hz) sensorineural hearing loss in the left ear. This acute hearing loss was a new finding compared with his prior audiograms, which had shown mild to moderate, age-related, sensorineural hearing loss bilaterally. Magnetic resonance imaging (MRI) of the brain found minimal nonspecific T2 hyperintensity signals in the midbrain and superior pons likely from small vessel ischemic disease, but imaging was otherwise unremarkable for internal auditory canal findings.

A preliminary diagnosis of acute idiopathic (presumed autoimmune-related) sensorineural hearing loss was made, and the patient was treated with a 60 mg dose of prednisone tapered over the course of 15 days. The steroid treatment led to the resolution of the patient's left eye pain, vestibular symptoms, and acute hearing loss. A repeat audiogram after corticosteroid treatment found a return to his chronic baseline. Two weeks after his last dose of steroids, however, the patient returned to his ophthalmologist for recurrent eye pain. The eye examination was unremarkable, but serum erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were mildly elevated at 29.0 mm/hr (normal range, 0–22.0 mm/hr) and 4.20 mg/dL (normal range, 1.0–3.0 mg/L) respectively, despite the 2-week course of steroid therapy the patient received before the laboratory testing.

The patient was referred to the Houston Methodist Hospital neuro-ophthalmology service. The neuro-ophthalmic examination was normal, but a left temporal artery biopsy revealed irregular intimal hyperplasia with focal segmental absence of the elastic lamina and CD-68 positive macrophages at the junction of the muscularis and the internal elastic lamina, compatible with healed or treated GCA (Fig. 1A–D). Serial repeat ESR and CRP tests were normal, and there was no recurrence of the hearing loss, vestibular symptoms, or eye pain off steroid therapy.

Discussion

AISHL can be fluctuating or progressive in nature and often presents unilaterally.[3,4] The symptoms typically develop acutely (less than 72 hours), which helps distinguish autoimmune hearing loss from the more common cause of hearing loss in the elderly, presbycusis.[3,4] AISHL affects 5 to 20 per 100 000 adults between the ages of 40 to 60 years annually.[3] Although up to 70% of patients recover some function spontaneously, AISHL is considered an otologic emergency requiring prompt referral and steroid treatment. AISHL, however, is a diagnosis of exclusion, and up to 16% of patients have an underlying autoimmune or neurologic etiology.[3] MRI with gadolinium is typically recommended for all patients with AISHL, but the potential need for other testing (including ESR and CRP) is less well known.[3]

Our report illustrates the case of a patient with GCA who presented with a clinical picture suggestive of a diagnosis of AISHL. Although many cases of GCA presenting with concomitant sensorineural hearing loss and vestibular symptoms have been reported in the literature, our case is unique in that our patient also presented with unilateral eye pain and tenderness. Table 1 summarizes the prior case reports of patients with GCA who presented with acute sensorineural hearing loss.[5–8] Clinicians should be aware that GCA can mimic AISHL in the elderly. Although AIHSL is predominantly unilateral, GCA should be included in the differential for elderly patients who present with acute, unilateral, or

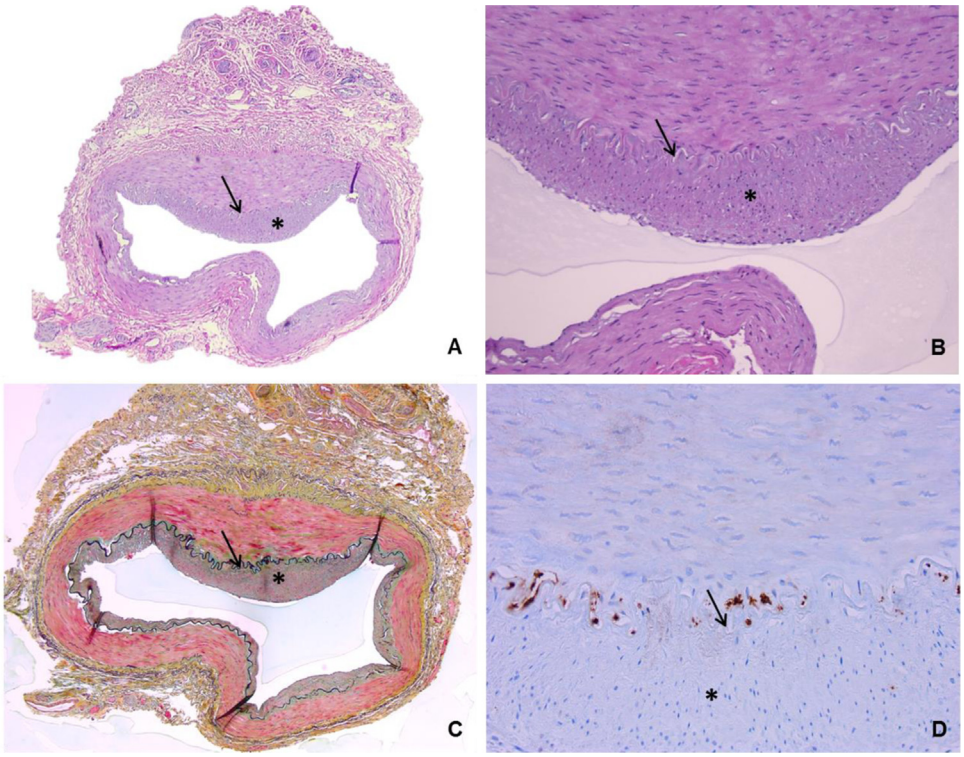

Fig. 1—Histopathologic findings in the temporal artery biopsy. (A) Low power view of the cross-section of the temporal artery highlights the irregular intimal hyperplasia (* = intima) and the absence of typical inflammatory infiltrate at the level of the elastic lamina (arrow) usually seen in active giant cell arteritis (GCA). Hematoxylin and eosin stain. Original magnification 4×. (B) Higher magnification demonstrates the irregular intimal hyperplasia adjacent to the internal elastic lamina (arrow). Hematoxylin and eosin stain. Original magnification 10×. (C) Movat stain that differentiates new fibrosis (in blue/green = *) from elastic lamina (black arrow) and muscularis layer (red/brown) shows that there is irregular elastic lamina with segmental absence of normal elastic lamina but with duplication of elastic lamina. The muscularis is thinned in some areas. Movat pentachrome stain. Original magnification 4×. (D) Macrophages seen in brown are present at the muscularis side of the elastic lamina (arrow) when labelled with immunohistochemistry. Immunohistochemistry using CD68 antibody (for macrophages) with 3,3'-Diaminobenzidine chromogen and hematoxylin counterstain (Ventana Automated Staining System, Oro Valley, Ariz.). Original magnification 20×.

bilateral sensorineural hearing loss. The presence of concomitant ocular symptoms should prompt consideration for an ESR, CRP, and a temporal artery biopsy in patients with presumed AISHL. Although both AISHL and GCA are treated with steroids, the markedly shorter duration and lower dose regimen used in the treatment of AISHL

Table 1—Summary of patients with GCA presenting with acute unilateral sensorineural hearing

Reference	Patient	Sex	Age (years)	Fever	Headache	Jaw Claudication	Temporal Artery Tenderness	Eye Pain or Tenderness	Visual Disturbance	Sensorineural Hearing Loss	Vertigo	Temporal Artery Biopsy Results
Francis and Boddie[5]	1	M	59	–	+	–	–	–	–	+	–	–
McKennan et al.[6]	2	F	84	–	–	–	–	–	+	+	+	+
McKennan et al.[6]	3	F	85	–	–	–	–	–	–	+	+	+
McKennan et al.[6]	4	F	79	–	+	–	+	–	–	+	+	+
Hausch and Harrington[7]	5	F	76	–	+	–	–	–	+	+	–	+
Hausch and Harrington[7]	6	F	76	–	–	+	+	–	–	+	–	+
Hausch and Harrington[7]	7	F	62	+	+	–	–	–	+	+	–	+
Hausch and Harrington[7]	8	F	59	+	+	–	+	–	–	+	–	+
Loffredo et al.[8]	9	F	80	–	+	–	–	–	–	+	–	+
Le et al. 2018	10	M	74	–	–	–	–	+	–	+	+	+

Age, sex, clinical features, and temporal artery biopsy results of 10 patients diagnosed with GCA who presented with sensorineural hearing loss. The data from patients 1–9 were obtained via a review of the literature (from 1982–2017), and patient 10 depicts information about our patient.[5–8]
The "+" symbol illustrates the presence of a symptom, and the "–" symbol illustrates the absence of such.
The table represents cases of GCA documented between 1982 and 2017 but does not report all cases of GCA presenting with bilateral hearing loss.

may be insufficient to treat GCA, can delay the diagnosis of GCA, and potentially cause irreversible blindness—the most feared complication of GCA.

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Multimodality imaging of multifocal ocular bartonellosis with an optic nerve head mass

A 26-year-old male patient presented with a 3-week history of sudden-onset painless loss of vision in his left eye. He reported a mild occipital headache that subsequently resolved without treatment over the course of 7 days but no other symptoms. There was no fever, lymphadenopathy, or other constitutional features, and the patient was otherwise physically well. There was no medical history or family history of note and no history of foreign travel. He did not take any regular medications.

On direct questioning, the patient admitted to being scratched by a partner's cat 2 weeks before to the onset of his visual loss.

On examination, the best-corrected visual acuity was 20/20 OD and 20/200 OS. Examination of the anterior segments, including the anterior chambers, was unremarkable. There were no preauricular and submandibular adenopathy. A left eye relative afferent pupillary defect (RAPD) was present. Fundal examination of the left eye showed swelling and a mass lesion of the left optic nerve head projecting into the vitreous. Fundal examination of the right eye did not reveal any abnormalities.

Multimodality retinal imaging, including color fundus photography, ocular angiography (fundus fluorescein angiography [FFA] and indocyanine green [ICG] angiography on HRA2 Heidelberg system), and Spectralis optical coherence tomography (SD-OCT), was acquired to investigate the case (Figs. 1–3).

Systemic investigations, including magnetic resonance imaging of the head and orbit, revealed no intracranial or orbital pathology and no compression of the optic nerve or optic chiasm. There was no abdominal mass. Chest x-ray was normal. Full blood count, urea and electrolytes, liver function tests, glucose, serum angiotensin-converting enzyme, and C-reactive protein were all unremarkable; however, erythrocyte sedimentation rate was high-normal (8 mm/hour). Serological tests showed negative *Toxoplasma gondii* IgM and IgG, *Borrelia burgdorferi*, and *Treponema pallidum* antibody assays, and a negative interferon-gamma (T-Spot-TB) test. Serological tests demonstrated the presence of *Bartonella* antibodies (>128), suggestive of recent infection, and this was confirmed on repeat testing. An autoimmune screen (antineutrophil cytoplasmic antibodies, antinuclear antibodies) and protein electrophoresis did not reveal any abnormalities.

The mass lesion emanating from the optic disc was initially thought to be suggestive of a neoplastic pathological process or possibly an angioma. Figures 1A and 1B show that the swelling was focal and not diffuse as would be expected in a disc that is swollen due to compression, for example.