Bilateral Sensorineural Hearing Loss as Clinical Manifestation of Giant Cell Arteritis: A Case Report.

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Learning Objectives

- Giant cell arteritis (GCA) is a systemic vasculitis that rarely involves the vestibulocochlear system.
- Rapid treatment with pulse dose steroids may prevent irreversible vestibulocochlear system ischemia and hearing loss.

Case

73-year-old woman with diagnosis of GCA, on prednisone 15 mg daily, and heart failure (HF), presented with right-sided frontoparietal headaches, and ongoing right-sided hearing loss for two weeks. She had a recent hospitalization due to decompensated HF and was treated with intravenous furosemide, followed by oral furosemide upon discharge.

Physical examination:
- Right-sided sensorineural hearing loss (SNHL)

Differential diagnosis:
Furosemide-induced ototoxicity
GCA-associated hearing loss
Acute idiopathic sensorineural hearing loss (AISHL)

Clinical features and management on admission:
- **Laboratory analysis:** elevated erythrocyte sedimentation rate (ESR) at 130 mm/h and C-reactive protein (CRP) at 304 mg/dl.
- **Magnetic resonance of the brain without contrast:** multiple foci of FLAIR hyperintensity in the supratentorial white matter, suggestive of vasculitis (Figure 1).
- **Management:** IV methylprednisone 1mg/kg/day for 5 days. Furosemide was stopped. She had partial improvement of her symptoms and was discharged home on prednisone taper.

Clinical evolution and new hospital admission:
- Two weeks later, despite discontinuation of furosemide and being on prednisone, she presented with sudden contralateral (left side) hearing loss and vertigo, left-sided headaches and jaw claudication.
- **Laboratory analysis:** persistent ESR and CRP elevation at 118 mm/h and 157 mg/dl, respectively.
- **Audiometry evaluation:** severe right SNHL and moderate left SNHL.

Case Continued

Management during readmission and follow-up:
- Three-day course of IV methylprednisolone 500 mg daily, followed by a six-month prednisone taper.
- At six-month follow-up, her headaches resolved, and ESR and CRP decreased up to 2 mm/h and 0.4 mg/dl, respectively.
- Unfortunately, hearing loss did not improve, and cochlear implants were indicated as definitive treatment of SNHL.

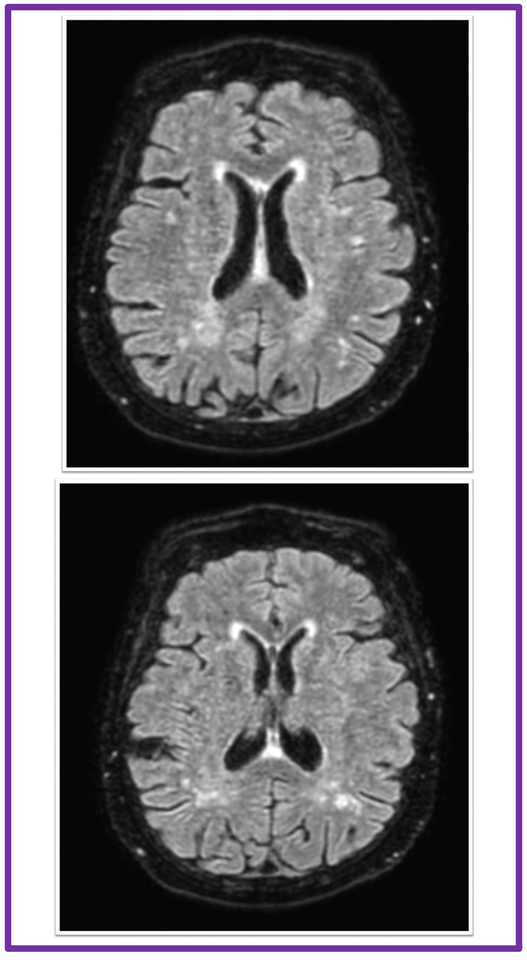

Figure 1. MRI of the brain w/o contrast with multiple foci of FLAIR hyperintensity in the supratentorial white matter, related to small vessel ischemic changes, suggestive of vasculitic process.

Discussion

- Only few similar case reports have described an association of GCA and SNHL. Most of these patients improved with early glucocorticoid therapy, as similarly observed in patients with AISHL.
- In the present case, a delay in establishing a definite diagnosis of GCA-associated hearing loss postponed the initiation of high-dose glucocorticoid therapy, likely leading to SNHL progression.
- Pulse steroid is the mainstay of treatment in GCA once neurovascular complications are established.
- The possible mechanism of deafness in GCA patients includes inflammatory involvement of the vertebrobasilar or cochleovestibular vessels, leading to vessel narrowing or occlusion, ischemia, and subsequently irreversible SNHL.
- In conclusion, early recognition of SNHL in GCA is an emergency, and rapid treatment with pulse dose glucocorticoids (even in the presence of confounding risk factors for hearing loss) may prevent irreversible vestibulocochlear system ischemia and subsequent hearing loss.

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