CLINICAL REPORTS

Acute hearing loss in giant cell arteritis

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Summary

A case of giant cell arteritis presenting with acute hearing loss which was reversed by corticosteroid therapy is described.

Introduction

The classical picture of giant cell arteritis, an elderly patient with headache and an inflamed temporal artery, is well known. It is also well recognized that in some patients sudden blindness may be the first sign of the disease (Healey and Wilske, 1977).

However, when these cardinal manifestations are absent or vague and the patient develops an uncommon symptom, the diagnosis is less likely to be considered. This case illustrates a rare yet important presentation of giant cell arteritis which has not been adequately reported in the literature.

Case history

A 59-year-old man gave a 4-week history of temporal-occipital headache. The pain was constant but of varying severity being aggravated by leaning forward and coughing. Simple analgesia had proved ineffective. No disturbance of vision had occurred but over the preceding 48 hr he had complained of a discomfort in both ears associated with increasing deafness. He looked unwell and was overtly depressed, complaining of profound lethargy, malaise and anorexia.

General examination was normal and his temporal and occipital arteries were non-tender with normal pulsation. Neurological examination revealed bilateral sensorineural deafness which deteriorated significantly over the subsequent 24 hr (Fig. 1).

Investigations: Hb 11.9 g/dl, WBC 11.5 × 10⁹/l, erythrocyte sedimentation rate 102 mm in the first hour, serum electrolytes normal, liver function tests mildly deranged. Alkaline phosphatase 199 u./l (normal 30–100), gamma glutamyltransferase > 400 u./l (5–55), 5-nucleotidase 25 u./l (3–17). Protein electrophoresis revealed a marked increase in alpha₁, alpha₂ and gamma globulin bands, serum immunoglobulins were normal.

Skull and chest radiology, a computerized axial tomographic brain scan and isotope liver scan all proved normal.

Despite a negative temporal artery biopsy a diagnosis of giant cell arteritis was made on established clinical criteria (Malmvall et al., 1976) and prednisolone therapy started at a dose of 80 mg daily. Within 24 hr his headache had gone and he felt his hearing had returned to normal. Marked improvement in the latter was confirmed by repeated audiometry performed during full suppression of disease activity (Fig. 2).

Discussion

Otolaryngological manifestations of giant cell arteritis are varied and include headache, temporal-occipital headache, ear discomfort, hearing loss and deafness (Hansen, 1973; Bovill, 1979; Grootaert, 1980). A bilateral sensorineural deafness may be the first sign of the disease (Healey and Wilske, 1977) and deafness is often accompanied by pain (McCombe, 1977). In some cases, hearing loss is the only symptom (Levy et al., 1980).

The diagnosis of giant cell arteritis may be delayed because the classical signs of headache, temporal-occipital headache, ear discomfort and deafness are absent or vague and often the patient presents with atypical symptoms such as anorexia and malaise (Levy et al., 1980). The diagnosis of giant cell arteritis has also been delayed because the disease may manifest itself by constitutional symptoms such as fever, malaise and anorexia (Malmvall et al., 1976; Levy et al., 1980).

The classical picture of giant cell arteritis includes headache, temporal-occipital headache, ear discomfort and deafness. The diagnosis of giant cell arteritis may be delayed because the classical signs of headache, temporal-occipital headache, ear discomfort and deafness are absent or vague and often the patient presents with atypical symptoms such as anorexia and malaise. The diagnosis of giant cell arteritis has also been delayed because the disease may manifest itself by constitutional symptoms such as fever, malaise and anorexia.
arteritis are infrequent but well documented. Patients may present with masticatory claudication, tongue pain or frank lingual infarction (Sofferman, 1980). Acute and potentially irreversible hearing loss however has received scant attention in the literature.

Deafness was noted in association with giant cell arteritis as early as 1946 when one of seven cases described developed unilateral followed by bilateral deafness which resolved spontaneously (Cooke et al. 1946). A more recent study found five cases out of a group of 68 patients suffering from giant cell arteritis with hearing loss. In each case audiograms revealed a bilateral sensorineural deafness and when these were repeated, following steroid therapy, no demonstrable improvement was seen (Malmvall and Bengtsson, 1978).

Rapidly progressive hearing loss as a presenting symptom in giant cell arteritis has been reported in one case previously (Cody, 1971). This patient also experienced the typical symptoms of bitemporal headaches, fever and weight loss, and also complained of tinnitus and intermittent vertigo. Audiograms again demonstrated a bilateral sensorineural hearing loss, and marginal improvement was seen following corticosteroid therapy. The mechanism of deafness in this condition is probably explained by arteritic involvement of the posterior circulation or terminal cochleovestibular vasculature (Sofferman, 1980). As in the visual complications of giant cell arteritis, its reversibility may depend upon the early introduction of treatment. This might also explain some of the conflicting reports in the literature regarding the response to steroids of deafness caused by the arteritic process. This case confirms an original report of acute and progressive hearing loss presenting as an early and prominent symptom in this disease. Its reversibility, following early steroid therapy, is clearly demonstrated and by calling attention to this it is hoped the index of suspicion of giant cell arteritis as a potentially reversible cause of acute hearing loss will be raised.

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References
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