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Clinical records

The otological manifestations of Wegener's granulomatosis

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Abstract
Three cases of Wegener's granulomatosis are described in which the common presenting symptoms were those of aural discomfort and discharge. The otological manifestations of this disease process are discussed and a review of the literature is presented.

Key words: Ear, Wegener's granulomatosis; Otitis media with effusion; Otitis media; Hearing loss, sensorineural; Vertigo

Introduction

Wegener's granulomatosis is a disease primarily of the respiratory and renal systems. It is characterized by necrotizing granulomas and vasculitis of arterioles and venules. The nasal cavity or paranasal sinuses are usually the site of the presenting lesion, but as the condition is a multisystem disease it can present in a variety of ways (Kornblutt *et al.*, 1980).

Otological involvement is reported as occurring in 38 per cent of all cases and may occasionally be the first and only sign of the disease (Nicklasson and Stangeland, 1982). It may affect the ear in a number of ways. Bradley (1983) reviewed the condition and described three differing modes of involvement; otitis media with effusion, acute otitis media and sensorineural hearing loss. Vertigo may also be the presenting symptom (Bennett and Staker, 1987).

The purpose of this paper is to present three cases of Wegener's granulomatosis in which the common presenting features were otalgia and otorrhoea and where each patient had a myringitis with middle ear involvement. A review of the otological findings in this systemic disease is also presented.

Case reports

Case 1

A 31-year-old female presented with a two-month history of otalgia and otorrhea. She also complained of mild nasal congestion, fatigue and weight loss.

Examination revealed a mucoid discharge in the left external auditory canal and a left-sided otitis media with effusion. She failed to respond to systemic antibiotics and a myringotomy with ventilating tube was performed.

The discharge persisted despite these measures and she complained of an increasing post-auricular pain. The patient was admitted for investigation. On admission, she was now complaining of a right-sided pain and discharge. She had a right middle ear effusion and anterior rhinoscopy revealed crusting in the left nasal cavity.

Pure tone audiometry demonstrated a sensorineural hearing loss in the left ear. Erythrocyte sedimentation rate (ESR) was noted to be 68 mm/min.

Biopsies of the nasal mucosa and tympanic membrane were performed. Forty-eight hours later she developed very severe post-auricular pain and a left partial facial paralysis.

Histology confirmed Wegener's granulomatosis in both biopsy specimens. She was commenced on cyclophosphamide. The facial weakness and post-auricular pain resolved rapidly and she has made a good recovery. ESR three months after commencement of treatment had fallen to 40 mm/h from a maximum of 80 mm/h. The sensorineural hearing loss has persisted.

Case 2

A 27-year-old female presented with a five-week history of right-sided facial pain. She also complained of a left-sided otorrhoea and some bloodstained nasal discharge.

Examination revealed a left acute otitis media, a right middle ear effusion and markedly enlarged right inferior turbinate. Her audiogram displayed a conductive hearing loss on the left side. X-rays of her paranasal sinuses were suggestive of maxillary sinusitis and she was treated with intravenous antibiotics. ESR at that time was 108 mm/h.

She then underwent a left antral lavage which resulted in the aspiration of copious amounts of pus from that sinus.

There was no improvement in the patient's condition and she had a right radical antrostomy performed. Histology was initially reported as being suggestive of tuberculosis. Wegener's granulomatosis was diagnosed on further review of the specimens despite the absence of vasculitis. She was treated with cyclophosphamide and prednisolone and made an excellent recovery. Two months after beginning therapy her hearing had reverted to normal and her ESR was 11 mm/h.

Case 3

A 32-year-old female presented with a three-week history of severe left-sided facial pain. This was accompanied by a right-sided otorrhoea and left nasal obstruction with an associated blood stained nasal discharge.

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TABLE I
ORGAN SYSTEM INVOLVEMENT IN WEGENER'S GRANULOMATOSIS (FROM NICKLASSON AND STANGELAND, 1982)

Organ system	Percentage involvement
Lungs	100%
Nasal cavity, paranasal sinuses	95%
Kidneys	85%
Joints	52%
Eyes	49%
Skin	44%
Ears	38%
Heart	23%
Nervous system	20%

Examination revealed a right acute otitis media with considerable debris in the external auditory canal. There was a left middle ear effusion. Anterior rhinoscopy revealed that both middle turbinates were markedly congested and there was mucopus in the nasal cavity.

X-rays suggested a left maxillary sinusitis and she was treated with intravenous antibiotics. Her ESR was noted to be 120 mm/h. She underwent bilateral lavage and a large amount of pus was aspirated from her left maxillary sinus. She was discharged. She was admitted five days later complaining of a severe left-sided facial pain.

A left radical antrostomy was performed with a biopsy of the right tympanic membrane.

Histology of the maxillary antral mucosa was reported as Wegener's granulomatosis. The tympanic membrane specimen demonstrated non-specific inflammatory changes. She was commenced on cyclophosphamide and prednisolone.

She is still having recurrent sinusitis but is otherwise well. ESR after three months of treatment was 10 mm/h.

Discussion

Wegener's granulomatosis is a rare disease. The triad of upper and lower respiratory tract with renal involvement is frequently described but the vasculitis may affect many organs (Hodges *et al.*, 1987).

Table I demonstrates the variety and frequency of organ involvement. The disease has a peak incidence at about 40 years of age with a slight male preponderance (Nicklasson and Stangeland, 1982).

The ear can be involved in a number of ways (Table II).

(1) Otitis media with effusion: this can occur either unilaterally or bilaterally caused by nasopharyngeal ulceration with eustachian tube obstruction (Bradley, 1983).

(2) Acute otitis media: this is caused by primary involvement by the disease of the middle ear and mastoid cavity causing granulomatous destruction and dissemination throughout the temporal bone (Bradley, 1983). Facial nerve palsy has been reported (Hugh-Powers, 1974). The extremely severe post-auricular pain is probably due to the vasculitic properties of the process but the exact aetiology is unknown.

The cause of otorrhoea has not been explained: post-mortem studies of the external auditory meatus have revealed numerous healthy ceruminous glands but also the presence of purulent keratotic material (Friedmann and Bauer, 1973). Th tympanic membrane in these studies was moderately thickened but intact. It was covered by cellular granulation tissue. There was no evidence of a tympanic membrane perforation in any of the cases we have reported but all cases had debris in the external auditory canal. Whether the aural discharge is as a consequence of myringitis in its own right or secondary to middle ear mucosal changes is not certain.

(3) Sensorineural hearing loss: the aetiology is unknown but a number of theories have been put forward including deposition of immune complexes in the cochlea, granulomatous compression of the cochlear nerve or vasculitis of the cochlear vessels (Bennett and Staker, 1987). This type of hearing loss is reversible with immunosuppressive therapy (Kempf, 1989).

(4) Vertigo: vestibular involvement is a distinctly unusual presentation in contrast to the cochlear signs. There are two principal aetiological theories:
   (a) Immune complex deposition in the vestibular portion but the point is made that the cochlear portion is more susceptible to this phenomenon.
   (b) It is a manifestation of central nervous system involvement caused by a polyneuritis.

TABLE II
OTOLOGICAL MANIFESTATIONS OF WEGENER'S GRANULOMATOSIS

Symptoms
Otitis media with effusion
Acute otitis media
Sensorineural hearing loss
Vertigo

Neither of these theories can explain the disparity between the incidence of vestibular and cochlear symptoms but the absence of vertigo may be due to central compensation.

Clinical suspicion is very important in that the diagnosis should be considered in patients with middle ear disease that fails to respond to conventional therapeutic measures (Bradley, 1983). Clinically the constitutional upset is out of all proportion to the local findings. The facial or post-auricular pain experienced by all three patients was very severe. The ESR is elevated in all patients and is a good parameter of disease activity (Bradley, 1983). Anti-neutrophil cytoplasmic antibodies (ANCA) are extremely specific for Wegener's granulomatosis and are being used for the diagnosis and monitoring of the disease. They can be positive in patients with other vasculitic diseases but this is considered quite rare (Nolle *et al.*, 1989). None of our three patients were tested for these antibodies as they had been attended to prior to the commercial availability of the test.

The biopsy must be adequate in size and depth. It should be taken from the nasal mucosa. The tympanic membrane biopsy has proven to be inconclusive. A complete or typical histological picture is not always essential for a diagnosis of Wegener's granulomatosis. Vasculitis may in fact be absent (Fienberg, 1981). If the clinical pattern is suggestive of the diagnosis a less typical histological picture can be acceptable.

The condition is fatal if untreated and the onset of renal involvement reduces the prognosis for recovery. Treatment with cyclophosphamide and prednisolone can lead to 95 per cent remission (Kornblutt *et al.*, 1980). Therefore, the correct early diagnosis and subsequent treatment can lead to a marked improvement in the patient's morbidity and survival.

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