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Early diagnosis of Wegener's granulomatosis presenting with facial nerve palsy

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Abstract

Wegener's granulomatosis is a multi-system disease characterized by granuloma formation and a necrotizing vasculitis. It classically presents with involvement of the upper and lower respiratory and renal systems. However locoregional disease is common and may include otological manifestations. Facial nerve palsy has been reported during the course of the disease process but it is extremely rare for it to be the presenting feature. Previously reported cases have involved a protracted diagnostic process including exploratory tympanotomy, mastoidectomy and facial nerve decompression. We report a case of Wegener's granulomatosis which presented with a facial nerve palsy. An early diagnosis was achieved by measurement of the erythrocyte sedimentation rate (ESR), followed by serological assay of cytoplasmic anti-neutrophil cytoplasmic antibody (cANCA). Histological examination of nasal mucosal biopsies confirmed granuloma formation consistent with a diagnosis of Wegener's granulomatosis. This allowed early treatment with immunosuppressive therapy and avoided unnecessary and potentially hazardous middle ear surgery.

Key words: Wegener's granulomatosis; Facial paralysis

Case report

A previously fit 65-year-old woman was admitted with a right-sided facial nerve palsy associated with a painful discharging right ear and bilateral hearing loss, all of three months duration. There was no previous history of otological disease or previous history of upper respiratory, chest or renal disease or diabetes. On examination the patient had a myringitis of the right tympanic membrane with granulation tissue present and evidence of a middle ear effusion. On the left side there was also a middle ear effusion. The patient had a complete right-sided lower motor neurone facial nerve palsy but no other cranial nerve palsies or neurological signs. Pure tone audiometry revealed a dead ear on the right side and a mixed deficit (air conduction 70 dB: bone conduction 30 dB) on the left side.

Investigations revealed a normal white cell count, blood glucose, urea and electrolytes and chest X-ray and examination of the urine revealed no microscopic haematuria. Significantly however the erythrocyte sedimentation rate (ESR) was raised at 92 mm/hr. Biopsy of the granulations on the right tympanic membrane revealed non-specific granulation tissue only with no evidence of malignancy or granuloma formation. CT scanning revealed soft tissue opacification of both middle ear clefts but no bony destruction. In addition there was soft tissue thickening of the maxillary sinuses (see Figure 1). In view of the raised ESR, a cANCA test was performed which revealed a positive titre of 1:80. Multiple biopsies were taken of the maxillary antra, lateral wall of the nose and the inferior turbinates. These revealed granuloma formation with evidence of necrosis and ulceration (Figure 2). Ziehl Nielson staining for acid and alcohol fast bacilli was negative and a diagnosis of Wegener's granulomatosis was therefore made.

In view of the diagnosis, surgical intervention was avoided and the management of the patient continued by the respiratory physicians. She was commenced on prednisolone 40 mg twice daily on alternate days and co-trimoxazole 960 mg daily. After six weeks of this regimen there has been a significant reduction in the severity of

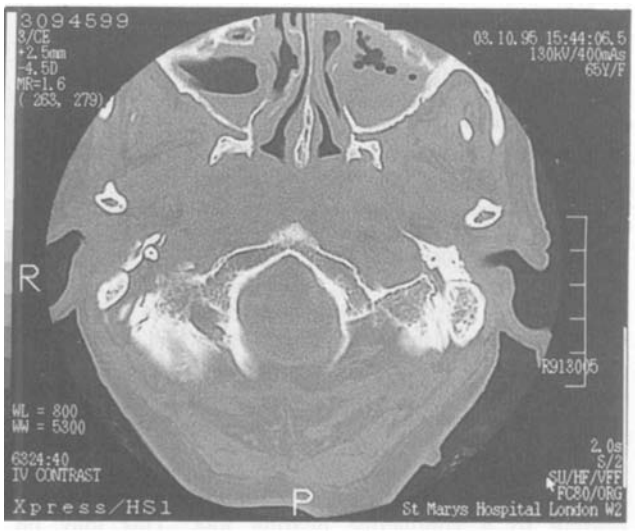

FIG. 1

Axial CT scan of the petrous temporal bones and paranasal sinuses showing soft tissue opacification of both middle ear clefts and soft tissue thickening within the maxillary sinuses.

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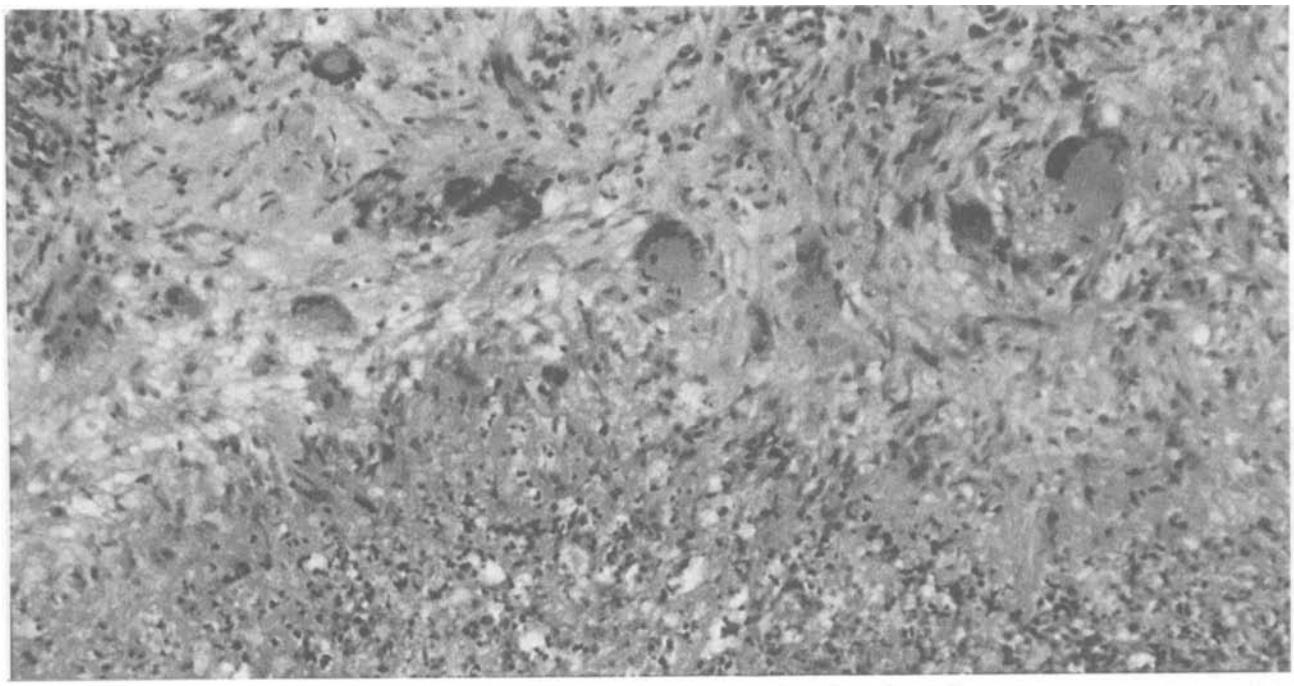

FIG. 2
Histological section of the nasal mucosa demonstrating numerous granulomata (H & E; ×250).

otalgia and otorrhoea and no evidence of progression to systemic disease. However, there has been only a slight improvement in the facial weakness and no alteration in hearing.

Discussion

Wegener's granulomatosis was first described in 1939 and classically comprises destructive lesions in the upper and lower respiratory tracts and glomerulonephritis. Histologically the lesions demonstrate granuloma formation and a necrotizing vasculitis. One-third of patients may present with a locoregional form of the disease which may last from months to years (Gross *et al.*, 1991), and may affect virtually any organ including the ear. Eventually these atypical cases progress to the classic generalized form of the disease which untreated has a mean survival of only five months (Fanci and Wolff, 1973). Therefore early diagnosis is important and is best achieved by a high index of clinical suspicion combined with measurement of the erythrocyte sedimentation rate. If this is raised, use should be made of the serological marker cANCA. In active systemic disease 95 per cent of patients have a raised titre of cANCA and 20–40 per cent have a raised titre for perinuclear anti-neutrophil cytoplasmic antibody (pANCA). In active locoregional disease 60 per cent of patients have a raised cANCA titre, which is therefore useful in the early diagnosis of locoregional otological Wegener's granulomatosis prior to the disease becoming systemic (Macias *et al.*, 1993).

Wegener's granulomatosis can affect the ear in a number of ways. A chronic middle ear effusion can occur leading to a conductive hearing loss. This may be secondary to granulation tissue in the middle ear cleft or from nasopharyngeal ulceration and obstruction of the Eustachian tube (Bradley, 1983). Occasionally the tympanic cavity may become filled with destructive masses of granulation tissue and may cause ossicular damage with erosion throughout the mastoid cavity and towards the petrous apex. Other otological manifestations include a sensorineural hearing loss which may be secondary to immune complex deposition within the cochlea, granulomatous compression of the cochlea nerve, or vasculitis of the cochlea vessels (Fenton and O'Sullivan, 1994). Vertigo may occur through similar mechanisms. Facial nerve palsy is a rare manifestation of Wegener's granulomatosis. The pathophysiology may be secondary to compression of the nerve in the temporal bone, particularly if the fallopian canal is dehiscent in the tympanic segment, or secondary to a vasculitis of the facial nerve microvasculature.

Facial nerve palsy may occur during the course of any granulomatous or vasculitic disease including tuberculosis, polyarteritis nodosa and sarcoidosis but very few cases have been reported in which a facial nerve palsy is the initial presenting feature of Wegener's granulomatosis. In a paper by Calonius and Christensen (1980) two cases were described in which hearing impairment and facial palsy were the initial signs of Wegener's granulomatosis. Patient A underwent a mastoidectomy and biopsy of the granulations revealed chronic inflammation and necrosis only. The facial nerve palsy was unchanged after the operation. Patient B underwent mastoidectomy and biopsy of the granulations revealed chronic non-specific inflammation only. Subsequently the patient underwent facial nerve decompression. Both procedures produced no improvement in the facial palsy. In patients with locoregional otological or classical Wegener's granulomatosis who subsequently develop facial nerve palsy, surgical intervention is equally unrewarding and some authors have suggested that it is contra-indicated since it aggravates the problem (Hugh Powers, 1974). In addition, biopsy of material from the tympanic membrane or middle ear often shows granulation tissue with chronic unspecific inflammation (Calonius and Christensen, 1980), and is therefore not diagnostic. However biopsy of the nasal mucosa is recommended (Fenton and O'Sullivan, 1994), as it is a technically simple procedure with a higher diagnostic yield.

Conclusion

Early diagnosis of Wegener's granulomatosis in patients presenting with locoregional otological disease and facial nerve palsy is possible by measurement of cANCA serology supplemented by biopsy of the nasal mucosa. This can lead to early administration of immunosuppressive therapy, which may delay or prevent the onset of the classical form of the disease and also improve the otological manifestations such as facial nerve palsy and sensorineural hearing loss (Macias *et al.*, 1993). It will also prevent unnecessary, difficult and potentially hazardous surgical intervention. It is important, therefore, that the clinician should retain a high index of clinical suspicion when managing patients with potential otological manifestations of Wegener's granulomatosis including facial nerve palsy. We suggest an ESR as a cheap and effective screening test in this situation and if raised, a cANCA test should be performed in order to aid the diagnosis.

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References

Bradley, P. J. (1983) Wegener's granulomatosis of the ear. *Journal of Laryngology and Otology* **97:** 623–626.

Calonius, I. H., Christensen, C. K. (1980) Hearing impairment and facial palsy as the initial signs of Wegener's granulomatosis. *Journal of Laryngology and Otology* **94:** 649–657.

Fanci, A. S., Wolff, S. M. (1973) Wegener's granulomatosis: studies in eighteen patients with a review of the literature. *Medicine* (Baltimore) **52:** 535–561.

Fenton, J. E., O'Sullivan, T. J. (1994) The otological manifestations of Wegener's granulomatosis. *Journal of Laryngology and Otology* **108:** 144-146.

Gross, W. L., Schmitt, W. H., Csernok, E. (1991) Antibeutrophil cytoplasmic autoantibody-associated diseases: a rheumatologist's perspective. *American Journal of Kidney Disease* **18:** 175–179.

Hugh Powers, W. (1974) Peripheral facial paralysis and systemic disease. *Otolaryngologic Clinics of North America* **7:** 397–404.

Macias, J. D., Wackym, P. A., McCabe, B. F. (1993) Early diagnosis of otological Wegener's granulomatosis using the serological marker c-ANCA. *Annals of Otology, Rhinology and Laryngology* **102:** 337–341.

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