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Wegener's granuloma – presenting as a unilateral parotid swelling

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Abstract

Wegener's granuloma is a systemic necrotizing vasculitis which predominantly affects the upper and lower respiratory tracts and kidneys. The nose and paranasal sinuses are often the first or main sites to be affected. Equally ear involvement is common and a recognized early manifestation of the disease. Salivary gland involvement is rare and is usually part of more widespread systemic involvement.

We present a 34-year-old female with a six-week history of hearing loss and a unilateral parotid swelling. One year previously she was diagnosed as having myalgic encephalomyelitis following a long period of post-viral malaise. The case points to the difficulties encountered when Wegener's granuloma presents in an unusual and varied way. A high index of suspicion coupled with early diagnosis and treatment is essential. The patient made a good recovery.

Key words: Parotid gland; Wegener's granulomatosis

Introduction

Wegener's granuloma is a systemic disease which primarily affects the upper and lower respiratory tracts and kidneys. It is a necrotizing, giant cell, granuloma characterized by the pathological changes of vasculitis from multinucleated giant cells. The sex incidence is equal and the peak age of onset is the fourth and fifth decades.

Investigations usually reveal a raised erythrocyte sedimentation rate (ESR), microscopic haematuria and multinodular or cavitating lesions on chest X-ray. Untreated the disease results in death from renal failure within a year. The mainstay of treatment is high doses of corticosteroids with the addition of the immunosuppressing agents cyclophosphamide and azathioprine. The prognosis has improved considerably in recent years.

Involvement of the salivary glands is extremely rare and is usually associated with systemic signs and symptoms of Wegener's granuloma. A patient is presented here in whom an ear infection and a unilateral parotid swelling were the initial manifestations. The diagnosis was made after careful clinical follow-up and laboratory investigations. The varied presentation led to a delay in diagnosis and points to the difficulties occasionally encountered when this condition presents in an unusual manner.

Case report

A 34-year-old female presented with a six-week history of pain, discharge and hearing loss from her right eye. She was previously diagnosed as having myalgic encephalomyelitis following a one-year history of post-viral malaise. Examination revealed an otitis externa, middle ear effusion and a right parotid swelling. The middle ear effusion was drained under local anaesthesia and topical antibiotic treatment commenced. Two weeks later her ear symptoms subsided and her main complaint was that of an enlarging lump around her left jaw (Figure 1). She also had haemoptysis and a bloody nasal discharge. On examination she was apyrexial with a large tender parotid swelling and marked trismus. The ENT examination was otherwise normal. Her haemoglobin was 10.1 and erythrocyte sedimentation rate (ESR) 41 with normal urea and electrolytes. Ward urine testing was normal as was the cytology and microbiology of the parotid lump.

A week's course of intravenous antibiotic led to no symptomatic improvement. A magnetic resonance imaging (MRI) scan (Figure 2) showed a diffuse swelling entirely within the parotid capsule with no evidence of abscess formation or malignancy. The parotid gland was explored and decompressed under general anaesthesia leading to marked improvement in symptoms and discharge from hospital. Histological biopsy showed features of chronic inflammation and the microbiology swab was sterile.

One week later the patient had worsening symptoms with bilateral parotid swellings, marked trismus, shortness of breath and aches and pains in joints of the upper and lower limbs. Her Hb was 9.1 and ESR 86.

An antineutrophil cytoplasmic antibody test (ANCA) was strongly positive and a chest X-ray showed bilateral diffuse infiltrates. A diagnosis of Wegener's systemic vasculitis was made. Treatment with steroids and cyclophosphamide led to a good recovery.

Discussion

Wegener's granuloma is a disease of unknown aetiology characterized by the presence of necrotizing granulomata and vasculitis. Commonly it involves the upper and lower respiratory tracts and the kidneys. Often the disease starts with severe rhinorrhoea with subsequent nasal mucosal ulceration followed by cough, haemoptysis and pleuritic pain. Skin and nervous system involvement may also accompany these symptoms (Murty, 1989).

Although ear involvement with Wegener's granuloma has been extensively described (Abraham-Inpijn, 1980; Nicklasson and Stangeland, 1982) salivary gland involvement is extremely rare. There are two recorded cases in the literature. In the first,

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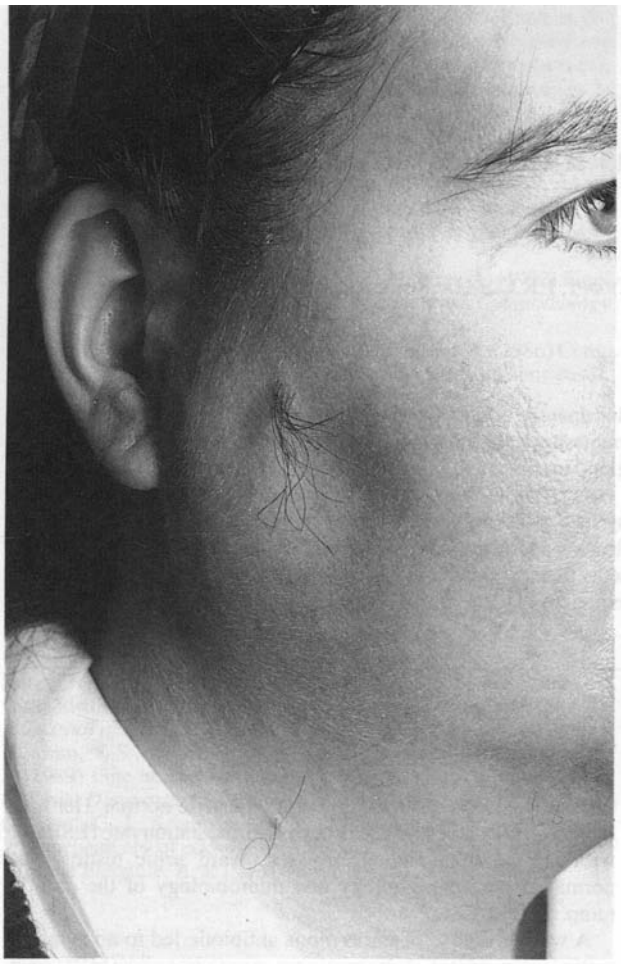

FIG. 1

Photograph at presentation showing diffuse left parotid swelling (patient's consent has been obtained).

Fauci *et al.* (1983) followed-up 85 patients for 21 years one of whom had granulomata of the parotid gland. In the second, Small *et al.* (1980) described a 59-year-old man who presented with a six-week history of bilateral painless swelling of the submandibular glands. The patient later developed relapsing polychondritis, serous otitis media, shortness of breath, cough and haemoptysis. A biopsy of the involved gland, chest X-ray and laboratory investigations were consistent with a diagnosis of Wegener's granuloma.

The standard description of Wegener's granuloma emphasizes respiratory and renal manifestations. However, the disease is truly systemic and the varied presentation may lead to a delay in diagnosis (Ridley *et al.*, 1988). A history of ear, nose and throat symptoms is common and in some studies (D'Cruz *et al.*, 1989) universal. In the same study all patients had systemic features of fever, malaise and weight loss with a mean duration of symptoms, before diagnosis, of 3.6 years.

The clinical presentation of the patient described in this report was diverse with features changing as the disease developed. The previous diagnosis of myalgic encephalomyelitis with its associated range of symptoms including malaise, fatigue, headaches and exhaustion meant that many of the ENT symptoms had to be elicited by direct questioning as the patient thought they were irrelevant to the present illness. Too little attention was paid to the initial presentation of a discharging deaf ear especially as it was later associated with haemoptysis.

Conclusions

Classical Wegener's granuloma should be diagnosed and treated promptly. Problems arise when patients have an unusual presentation and in these patients keeping a high index of suspicion is desirable. With early diagnosis and treatment the prognosis is excellent.

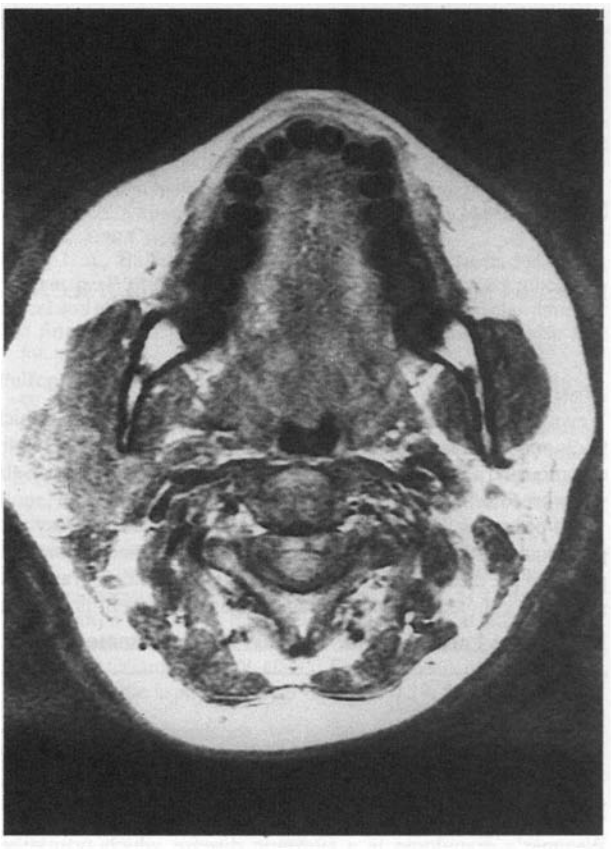

FIG. 2

MRI scan of left parotid gland showing diffuse swelling with no evidence of malignancy or abscess formation.

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