WEGENER'S GRANULOMATOSIS WITH ORAL LESIONS AT PRESENTATION

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ABSTRACT

It has long been appreciated that the mouth is the mirror for the rest of the body. Many systemic diseases have oral symptoms, which often coincide with widespread evidence of disease. In some cases, however the oral symptoms may be the presenting sign of systemic disease. We report a case of Wegener's Granulomatosis presented initially as oral lesions.

INTRODUCTION

Wegener's granulomatosis is a form of primary systemic vasculitis characterised by involvement of the lungs and upper airways and the kidneys[1]. Histological examination of the affected tissue demonstrates a necrotizing granulomatous vasculitis of the lungs together with focal proliferative glomerulonephritis.

Oral mucosal lesions were the presenting features found in 5% of patients in a recent study[2]. Although oral ulceration often occurs late in the disease, the hyperplastic gingivitis is thought to be an early feature and a series of biopsies may be required to obtain diagnostic tissue[3].

CASE REPORT

A fifty years old Malay man presented to the dentist with a history of painless gingival swelling, of more than one-month duration. This swelling tends to bleed whenever he brushes his teeth. On visual examination, the lesion was described as "discrete firm mass of tissue, toad -skin like, haemorrhagic in appearance and followed the contours of gingiva from the upper left to the upper right canine and was non-tender". Biopsy confirmed the lesion to be Wegener's granulomatosis.

One week after the onset of gingival swelling, he developed a left ear watery discharged, which subsided after treatment. He had history of fever for one week, associated with productive cough. On physical examination, he was found to be febrile. Minimum gum hypertrophy of the upper gingival was also observed. The left external auditory canal was inflamed but with intact typmpanic membrane. There were persistent coarse crepitations in the left lower lung zone posteriorly. Review of the other systems showed no obvious abnormality.

An initial chest X-ray done (Fig.1) showed a large nodular lesion in the posterior segment of the right upper lobe. High Resolution Computed Tomography (Fig. 2), showed non-cavitating nodules in the apical and posterior segments of the right upper lobe, apical and medial segments of the right lower lobe and both hilar regions (Fig.3). Computed Tomography (CT) of paranasal sinuses (Fig. 4) showed a soft tissue mass in the left frontal and ethmoidal, right sphenoid and both maxillary sinuses. No bony destruction was seen.

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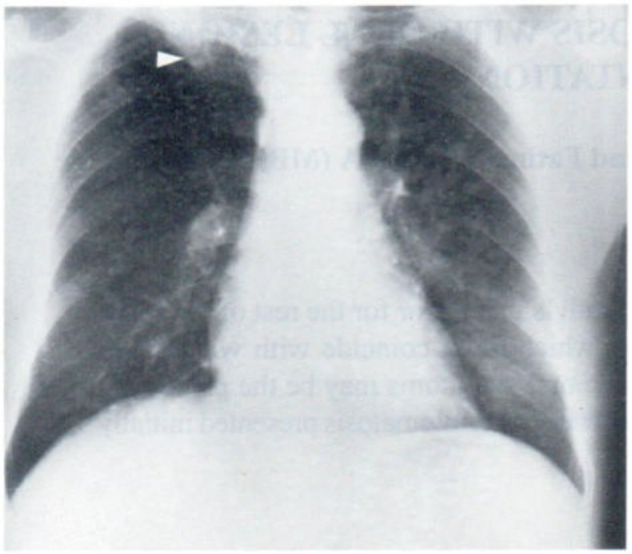

Fig. 1. Chest x-ray (PA view) showing a large nodular lesion in the posterior segment of the right upper lobe.

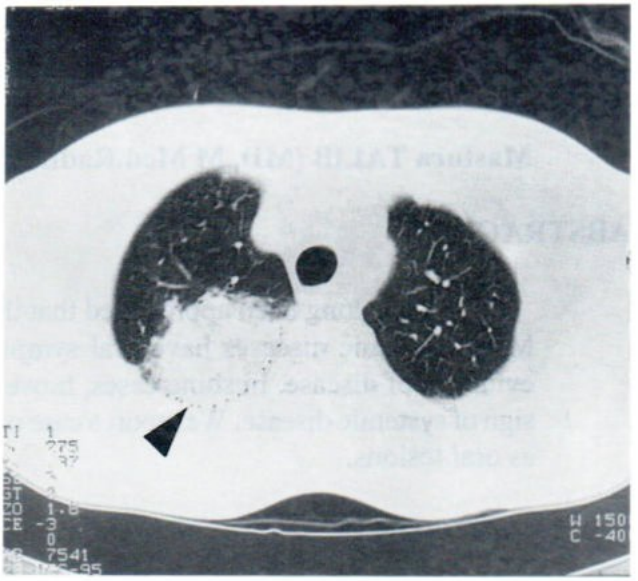

Fig. 2. HRCT of the chest demonstrating a large non-cavitating opacity in the apical segment of the right upper lobe.

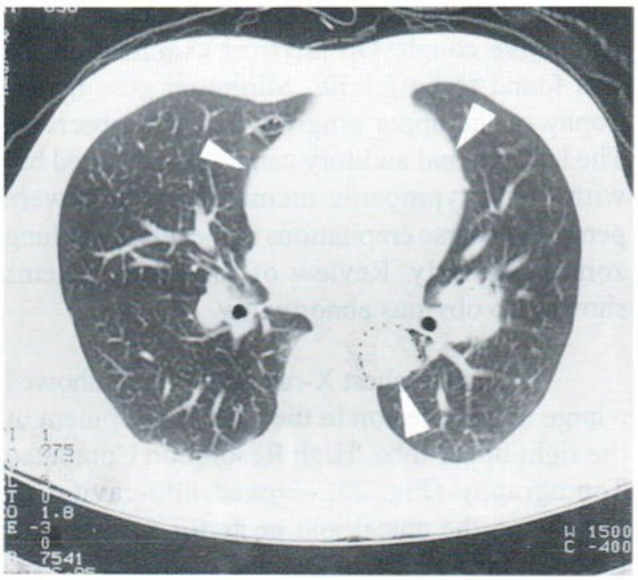

Fig. 3. Nodules seen at both hilar region on high resolution computer Tomography (HRCT)

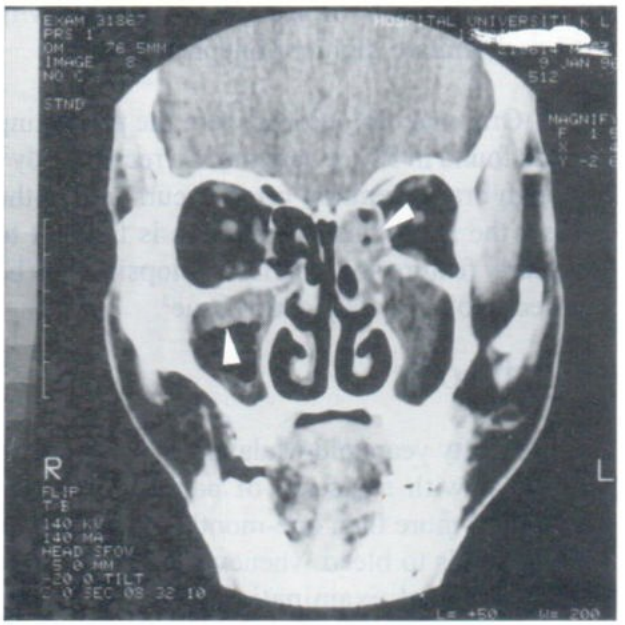

Fig. 4. Mucosal thickening of the left ethmoid and right maxillary sinus seen on CT.

CT guided percutaneous needle biopsy was performed for the right apical lung nodule. The histology showed strips of fibrous tissue infiltrated by mainly chronic inflammatory cells like lymphocytes and plasma cell. Vague granulomas with occasional multi-nucleated giant cells were seen clustering around vessels. Focal areas of necrosis were also present. The infiltrate in the tissue composed mainly of T-lymphocytes. These features were compatible with Wegener's granulomatosis.

Ultrasound of the abdomen was normal. He was treated with Cyclophosphamide 100mg daily and Prednisolone 30mg daily and is on follow up presently.

DISCUSSION

Wegener's granulornatosis is a form of primary systemic vasculitis, which affects predominantly small vessels and is associated with the presence of granulomata. In the localised form, the involvement is usually confined to the upper and lower respiratory tracts or both. In the commoner generalised form, the kidneys are also involved[1].

The classical presenting features are referable to the upper respiratory tracts. These may include rhinorhea, paranasal sinus pain and discharge and frequently with nasal mucosal symptoms.

Pulmonary infiltrations may take many forms. Radiologically, the lesion may be solitary or multiple nodular densities or infiltrations. They can have vague borders or be sharply circumscribed. The size of opacities varies greatly from less than one centimetre to more than nine centimetres. The infiltrations are frequently bilateral with no characteristic lobar distribution and cavitations are frequent[1].

Of particularly importance is that, infiltrations can be fleeting and transient. Focal areas of atelectasis adjacent to the infiltrated areas, as well as small pleural effusions, are infrequently seen. Mediastinal lymph node enlargement is extremely rare and calcification is not a common feature[1].

On computed tomography (CT), the classical radiological feature of Wegener's granulomatosis is the presence of multiple pulmonary nodules, frequently cavitating. Studies which have attempted to correlate the radiological and histological findings in patients with Wegener's granulomatosis have suggested that the nodules are usually found in close proximity to pulmonary vessels involved by the arteritis.

Wegener described oral ulceration at necropsy in most of his original cases, but regarded it as a late manifestation of the disease.

One of the diagnostic criteria for Wegener's granulomatosis is oral ulceration as defined by the American College of Rheumatology[4], although oral ulceration often occurs late in the disease, gingival hyperplasia is thought to be an early feature and a series of biopsies may be required to obtain diagnostic tissue[3].

Other oral lesions include palatal ulceration, lingual ulcer, aphthous ulcer, non-healing extraction sockets and gingivitis, which have been infrequently described. Wegener's granulomatosis presents in the early stage as characteristic hyperplastic gingivitis, named by authors as 'strawberry gum', which fails to respond to conventional periodontal therapy.

A case has been reported, in which this clinically distinctive gingivitis was the presenting lesion with serous otitis, hence illustrating that frequently occurring entities such as Weneger's granulomatosis should be considered in the differential diagnosis of localised gingival lesions

which fails to respond to conventional therapy[5].

Middle ear involvement is often encountered in association with the nasopharyngeal and sinus disease[6]. Indeed serous otitis media may be the earliest presenting feature[7]. Severe paranasal sinusitis, nasopharyngeal ulceration with nasal septa perforation may be a feature. In the order of frequency, the maxillary, ethmoidal, frontal followed by sphenoid sinuses are involved, although pansinusitis is quite common[3].

Air-fluid levels seen on paranasal sinus roentgenograms indicate secondary bacterial infection. When the primary sinus disease causes severe damage to the sinus mucosal surfaces, the control of secondary bacterial infection becomes quite difficult probably because of impaired drainage mechanism.

In conclusion, this is a good example of Wegener's granulomatosis presenting as gingival hyperplasia and hence biopsy of such lesion should be done for diagnosis and treatment purposes.

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