

# An Unusual Case of Wegener's Granulomatosis

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## Summary

A 35-year-old Malay man underwent treatment for uveitis of the right eye in 1992 but developed marked visual impairment in the affected eye after he failed to attend follow-up. Two years later, he complained of difficulty swallowing and was found to have left sided X and XI cranial nerve palsies. Chest radiograph showed a cavitating lesion in the lower zone of the right lung field. Inflammation and perforation of the nasal septum was found on examination of his upper respiratory tract. Punch biopsies taken from that area showed chronic inflammatory change and necrotizing vasculitides. The patient was diagnosed as having Wegener's granulomatosis and made a very good recovery with immunosuppressive therapy.

**Key Words:** Uveitis, Vasculitis

## Introduction

In Malaysia, Wegener's Granulomatosis (WG) is extremely rare and there have been no cases reported in the literature so far. The clinical progression (untreated) in our patient is also intriguing, particularly the development of cranial nerve palsies which is found only in a minority of patients with WG<sup>1</sup>.

## Case Report

Our patient first presented to the Eye Unit with a pain and redness of his right eye. He was subsequently diagnosed to have uveitis and was commenced on topical corticosteroids. However, the patient failed to attend for follow up therapy and subsequently became blind in that eye.

Further problems gradually arose over the next 2 years. In that time, he had twice sought treatment at another hospital for recurrent nasal discharge which had been noted to be foul smelling. He had also been troubled by cough for a year and weight loss for 6 months. There was also a history of haematuria on 2 occasions. Finally, he was admitted to hospital with a 5 day history of hoarseness of voice, dysphagia and nasal regurgitation of fluid.

Examination showed a cachexic patient who had a left vocal cord palsy and wasting of the left sternocleidomastoid muscle with intact palatal sensation, thus suggesting a X and XI nerve palsy. The nasal septum had raw inflamed areas and was perforated. A chest radiograph (CXR) showed a cavitating lesion in the right lower zone (Figure 1), but the computerized tomography (CT) scan of the brain showed no specific abnormalities. Blood investigations yielded a raised erythrocyte sedimentation rate (ESR) of 75 mm/hour and a repeatedly positive rheumatoid factor. No haematuria or albuminuria was present on examination of the urine and a fiberoptic bronchoscopy did not yield any significant findings.

Biopsies of the nasal septum were taken and histological examination showed chronic inflammatory changes with plasma cell infiltrates and areas of necrotizing vasculitis. Although we were not able to obtain results for the anti-neutrophil cytoplasmic antibody test (ANCA), the nasal and pulmonary involvement, coupled with the presence of necrotizing vasculitis, was sufficient for us to make the diagnosis of WG<sup>1,2</sup>.

The patient was started on oral cyclophosphamide and

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prednisolone and rapidly improved on this treatment. There was resolution of the cavity on the CXR and our patient gained 10kg in weight within a few months. The ESR gradually fell to normal levels. After 2 months therapy, thrombocytopenia and haematuria was noted but this resolved when azathioprine was used in place of the cyclophosphamide.

After 6 months of therapy the patient was unfortunately lost to our follow up. He had improved so much that he was now working full time at an electronics factory on the other side of the country, and had decided to continue his treatment at a hospital closer to his workplace.

### Discussion

This patient may well be the first case of WG reported in Malaysia, as there were certainly no reports from Malaysia found in the literature in a MEDLINE search from 1966 till present. In fact, WG appears to be a rare entity in Asia - in a Hong Kong study, no cases

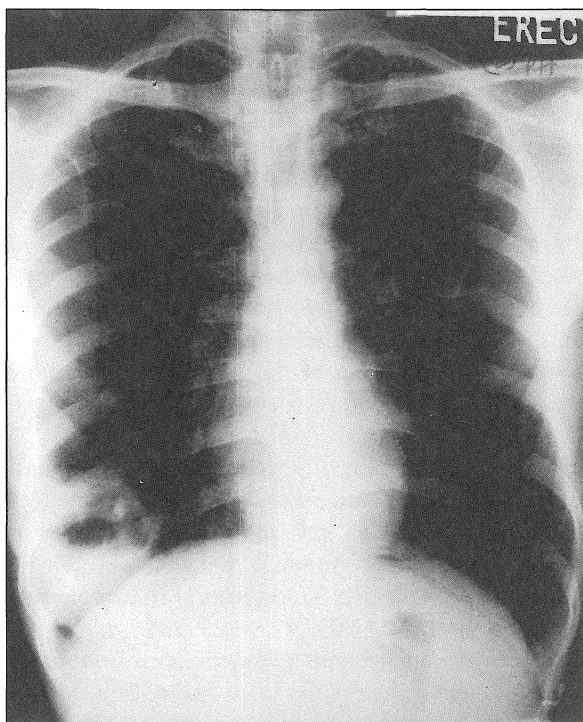
of WG existed in a group of 42 vasculitic patients with positive ANCA<sup>3</sup>.

WG belongs to the family of diseases with necrotizing vasculitis, which include entities such as polyarteritis nodosa, Churg Strauss syndrome and microscopic polyangiitis. There are no definite diagnostic criteria for WG but in the largest published series of WG patients from the National Institute of Health (NIH), America, cases were defined on the basis of a compatible clinical history plus histopathologic evidence of either vasculitis or granulomatous changes in a typical organ system<sup>1</sup>. It was also noted that the relatively small biopsy specimens obtained from the head and neck areas of patients with WG seldom showed the typical granulomatous lesions and our experience certainly reflects that<sup>1</sup>. Specimens from the kidney and open lung biopsies were better able to demonstrate such granulomas.

The NIH series also showed that the mean period of time from onset of symptoms to diagnosis of WG was 15 months and that some patients did have relatively indolent courses. Although only 18% of patients initially presented with glomerulonephritis, 77% went on to develop renal involvement within the first 2 years. Unfortunately, although our patient gave a history of haematuria even before the diagnosis of WG was suspected, his urine analysis was negative for albumin or red cells at the time we saw him.

Our patient illustrates well the multi system involvement that can be seen in WG. He initially presented with ocular involvement, then developed nasal symptoms and finally had pulmonary and central nervous system (CNS) features. In the NIH series, only 8% of the patients had CNS manifestations, and our patient certainly falls into this rare group. A positive Rheumatoid Factor, as seen in our patient, is a more common finding which occurs in 30-40% of patients with WG.

Prednisolone and low dose daily cyclophosphamide have been prospectively shown in the NIH series to be effective treatments of WG and our patient responded very well to this therapy. However, this regime is not free from side effects with cystitis being one of the most commonly noted problems<sup>1</sup>.



**Fig. 1:** Chest radiograph showing a cavitating lesion in the lower zone of the right lung field

Haematuria was noted in our patient during cyclophosphamide therapy and but this resolved when the drug was discontinued. Fortunately, he was then already in remission and continued to do well when put on Azathioprine instead. Nonetheless, we must bear in mind that the NIH series showed a 50% relapse rate among those in remission and it is our hope that our patient will continue with this follow-up and treatment wherever he may be.

This case of WG may well be the first of its kind

reported in Malaysia and provides a fascinating insight into the natural history of this disorder if left undiagnosed, particularly with regards to the complex multi system involvement that can slowly but progressively develop.

### Acknowledgements

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## Typhoid Thyroiditis

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### Summary

Acute suppurative thyroiditis in a 62 year old lady with enteric fever is reported. Plain radiography of the neck showed a distinct localised abscess cavity with air fluid level. A rare causative agent *Salmonella typhi* was isolated. Needle aspiration and antibiotics resulted in complete recovery.

**Key Words:** Acute suppurative thyroiditis, Thyroid abscess, Enteric fever, Salmonella infection, Typhoid fever