

Refractory Wegener's granulomatosis presenting with alveolar haemorrhage, treated with rituximab

Joana Caetano,¹ Marisa Fernandes das Neves,^{1,2} Susana Oliveira,¹ José Delgado Alves^{1,2}

¹Department of Medicine 4, Fernando Fonseca Hospital, Amadora, Portugal
²CEDOC – Center for Chronic Diseases of NOVA Medical School, Lisbon, Portugal

Correspondence to
 Professor José Delgado Alves,
 jose.alves@fcm.unl.pt

Accepted 28 November 2014

DESCRIPTION

A previously healthy 57-year-old man presented with epistaxis, crusted nasal ulcers and weight loss. CT scan of the sinuses revealed an inflammatory swelling of the ethmoid and maxillary sinuses (figure 1) and CT scan of the lungs showed two pulmonary nodular lesions in both upper lobes. Laboratory investigation showed an elevated erythrocyte sedimentation rate (97 mm/h) and a positive anti-PR3-antineutrophil cytoplasmic antibody.

The diagnosis of Wegener's granulomatosis (WG) was made and the patient was started on immunosuppression with cyclophosphamide and glucocorticoids followed by maintenance therapy with azathioprine, with subsequent disease remission.

After 2 years, the disease relapsed with haemoptysis and external popliteal sciatic mononeuropathy. A thoracic CT scan revealed multiple pulmonary nodular lesions and ground-glass infiltrate of the right lower lobe, compatible with alveolar haemorrhage (figure 2). The patient started treatment with rituximab with completely resolution of the symptoms and pulmonary lesions. He remained asymptomatic at 2-year follow-up.

WG is a primary systemic vasculitis, with predilection for the respiratory tract and kidney involvement. Although pulmonary findings are common, diffuse alveolar haemorrhage is a rare and life-threatening manifestation of WG. It is associated with six times greater mortality than vasculitis without pulmonary haemorrhage.¹

Cyclophosphamide is still the standard induction therapy for patients with WG. Nevertheless, relapse

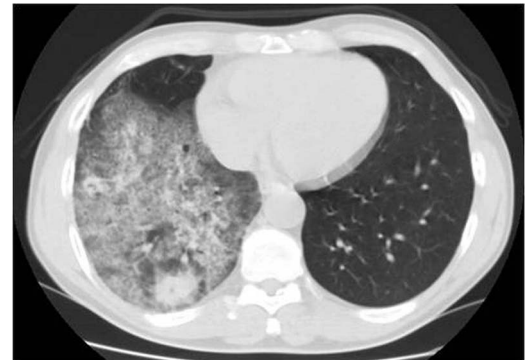


Figure 2 Thoracic CT scan showing pulmonary nodular lesions and alveolar haemorrhage in the right lower lobe.

is common in up to 50% of patients and rituximab has been proving to be an effective remission inducing agent in severe refractory WG.^{1 2}

Learning points

- ▶ Despite the poor outcome of patients with alveolar haemorrhage, it can have a benign course with prompt immunosuppression.
- ▶ Rituximab is an effective immunosuppressive therapy for the induction and maintenance of remission in refractory Wegener's granulomatosis.
- ▶ As most patients with antineutrophil cytoplasmic antibody associated-vasculitis relapse, a close monitoring of the patients is crucial for an early diagnosis and therapeutics.

Contributors JC and MFN participated in drafting of the manuscript. JDA and SO revised it critically for important intellectual content. All the authors contributed to conception of the work and acquisition of data.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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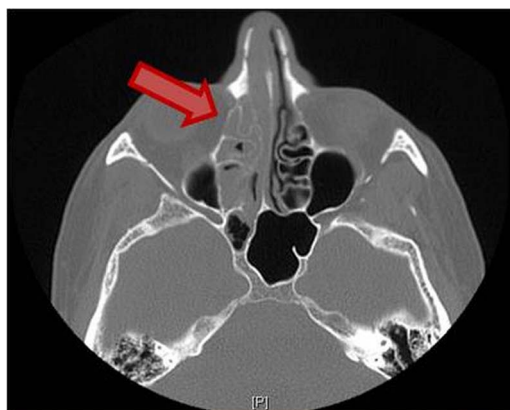


Figure 1 CT scan of the sinuses showing an inflammatory swelling of the ethmoid and maxillary sinuses (arrow).



To cite: Caetano J, Fernandes das Neves M, Oliveira S, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-208510

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