

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

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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.

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Competing interests None.

Patient consent Obtained.

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



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