Hoarseness of voice as the herald of granulomatosis with polyangiitis

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DESCRIPTION

A previously healthy 15-year-old boy presented with hoarseness of voice and progressive dyspnoea. A nodule was found over his vocal cord which was excised. Unfortunately, histopathology was inconclusive. The diagnosis was presumed to be allergic laryngitis and he was managed with oral antihistamines and intramuscular hydrocortisone with a good response. Six weeks later he felt unwell with deteriorating dyspnoea, productive cough with blood-tinged sputum and fever. He also reported decreasing urine output that was tea coloured. On presentation vitals were stable except for low-grade fever. He was in moderate distress with tachypnoea. There was no skin rash or arthralgia. History was negative for bronchial asthma. Previously serum creatinine was normal and it had sharply risen to 700 µmol/L within a few weeks; renal ultrasound was unremarkable. He had severe metabolic acidosis with a pH of 7.04; urinalysis revealed 50 red blood cells/high power field and protein-creatinine

ratio was 0.87. HIV, hepatitis B and C viral serology was negative. ANA (antinuclear antibody), antiglomerular basement membrane (GBM) antibody and perinuclear antineutrophil cytoplasmic antibodies (P-ANCA) were negative. Complement C3 and C4 were normal. Cytoplasmic ANCA (C-ANCA) (PR3) titre of 32 U/mL (range 0–10) was the only positive autoimmune marker. Chest X-ray showed multiple nodular lesions and chest CT revealed scattered pulmonary nodules with central necrosis (figure 1). Renal biopsy revealed pauci-immune necrotising and crescentic glomerulonephritis. Crescents were all cellular and fibrinoid necrosis was present (figure 2). Based on the clinical, biochemical, histopathological findings, the diagnosis was granulomatosis with polyangiitis (GPA) (vasculitis, granulomas, no asthma). Differential diagnoses include: pauci-immune glomerulonephritis (PIGN) (renal limited, no systemic vasculitis, P-ANCA (myeloperoxidase, MPO) positive); microscopic polyangiitis (MPA) (vasculitis with no asthma or

Figure 1 Chest X-ray showing scattered patchy infiltrates in all lung fields. Chest CT showing multiple scattered lung nodules variable in size, some with cavitation.

Figure 2 Renal biopsy showing a glomerulus with cellular crescent and fibrinoid necrosis—from left to right (H&E stain; Jones Silver stain; Trichrome stain) all at 200×.

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Learning points

- ▶ In granulomatosis with polyangiitis (GPA) subglottic stenosis occurs in 16–20% of all patients and in up to 50% of paediatric patients representing the most characteristic and serious laryngeal lesion. This manifests clinically with a hoarse voice.²
- ▶ The most commonly reported chest X-ray findings in GPA are nodules (20–90%) and patchy or diffuse opacities (20–50%).³
- ▶ The severity of the renal biopsy findings in GPA and microscopic polyangiitis generally correspond to the severity of the clinical presentation. In patients with asymptomatic haematuria the lesion present is mild focal and segmental glomerulonephritis in contrast to a diffuse necrotising and crescentic glomerulonephritis in patients with acute kidney injury.⁴

granulomas) and eosinophilic granulomatosis with polyangiitis (EGPA) (eosinophilia, vasculitis, asthma and granulomas).¹ Renal histopathology of GPA, PIGN, MPA and EGPA all have

the same pattern of injury. Based on the diagnosis and clinical presentation, pulse steroid was administered followed by oral prednisolone 1 mg/kg/day. Thereafter, our patient underwent seven sessions of therapeutic plasma exchange followed by oral cyclophosphamide 1 mg/kg daily. Six weeks following treatment he had improved clinically with regard to his respiratory symptoms but unfortunately did not regain renal function and remains dialysis dependent.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Jennette JC, Falk RJ, Bacon PA, *et al.* 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. *Arthritis Rheum* 2013;65:1–11.
- 2 Gluth MB, Shinnars PA, Kasperbauer JL. Subglottic stenosis associated with Wegener's granulomatosis. *Laryngoscope* 2003;113:1304–7.
- 3 Lee KS, Kim TS, Fujimoto K, *et al.* Thoracic manifestation of Wegener's granulomatosis: CT findings in 30 patients. *Eur Radiol* 2003;13:43–51.
- 4 Fauci AS, Haynes BF, Katz P, *et al.* Wegener's granulomatosis: prospective clinical and therapeutic experience with 85 patients for 21 years. *Ann Intern Med* 1983;98:76–85.

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