EMERGENCY CASEBOOK

Fulminant Wegener's granulomatosis presenting as epistaxis

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The case of a 63-year-old woman who presented to the emergency department with epistaxis and haemodynamic instability is reported. Subsequent investigation showed renal failure and multiple pulmonary nodules. A positive proteinase 3 antineutrophil cytoplasmic antibody test supported the diagnosis of fulminant Wegener's granulomatosis, requiring urgent dialysis, plasma exchange and immunosuppression. This is the first report in the emergency literature of Wegener's granulomatosis presenting as acute epistaxis. Emergency physicians should consider Wegener's granulomatosis in patients with atypical epistaxis. In patients presenting with clinically severe, active disease early proteinase 3 antineutrophil cytoplasmic antibody testing is recommended.

A 63-year-old Caucasian woman presented to the emergency department with spontaneous unilateral epistaxis of 12 h duration. She gave a history of exertional dyspnoea, haemoptysis and 4 kg weight loss over 3 months. Medical history included bilateral sudden sensorineural deafness. She was referred to the on-call ENT team.

On assessment, her pulse was 120 beats/min, blood pressure 156/80 mm Hg lying, 126/74 mm Hg standing, O₂ saturation 97% on air, respiratory rate 20 breaths/min and temperature 36.3°C. Rhinoscopy showed nasal crusting and a large haematoma overlying Little's area. This was removed and a friable area underneath cauterised with silver nitrate.

In view of the haemodynamic instability, a full blood count, coagulation and renal profile were requested. Laboratory findings showed a normocytic anaemia with haemoglobin 63 g/l, white cell count $8.5 \times 10^9/l$, C reactive protein 234 mg/l, creatinine 935 $\mu\text{mol/l}$ and potassium 6.0 mmol/l. Coagulation studies were normal. Urine analysis was requested, and showed microscopic haematuria.

The patient was transfused with three units of cross-matched blood, and plasma potassium levels were controlled with an insulin-dextrose infusion. Intravenous cefuroxime was commenced and the patient's fluid restricted to 1 litre/24 h in view of her renal function. Urgent ultrasound of the abdomen was performed, which showed no evidence of hydronephrosis or a renal lesion.

A review of her medical records showed that she was under investigation by a respiratory physician for haemoptysis. A recent CT scan of the thorax (fig 1) had shown a 2 cm mass in the right upper lobe and nodules of similar size throughout both lung fields, which had not responded to a month's course of ciprofloxacin.

In view of the triad of haematuria, lung nodules and epistaxis, a proteinase 3 antineutrophil cytoplasmic antibody (PR3 ANCA) determination was requested. This was positive at 615 U/ml (normal <1), strongly supporting the diagnosis of Wegener's granulomatosis. The patient was transferred for dialysis, plasma exchange and cyclophosphamide treatment. A renal biopsy was not performed, in accordance with the patient's wishes. Her renal function improved, but unfortu-

nately suffered a spontaneous intracerebral haemorrhage, which despite neurosurgical intervention led to her death 2 weeks later.

DISCUSSION

Wegener's granulomatosis (described by the German pathologist Friedrich Wegener in 1939)¹ is a multisystem granulomatous condition of unknown aetiology. It is defined by the American College of Rheumatology as the presence of two of four clinopathological criteria² (box).

ENT problems are found in 80% of patients and give the main clue to the diagnosis.³ Those that may present in the emergency department include stridor from subglottic stenosis, oral ulceration, sinusitis, otitis media and sudden conductive or sensorineural deafness.⁴ Necrosis of vessels in the cartilaginous septum leads to epistaxis, septal perforation and eventually a saddle-nose deformity.

Depending on the history, an ENT examination and chest radiograph or urine analysis should be performed. If characteristic abnormalities are found, the next step is either nasal or renal biopsy (the gold standard) or PR3 ANCA testing. In Wegener's granulomatosis, autoantibodies are directed at proteinase 3, a proteinase found in the granules of neutrophils. Meta-analysis has shown the sensitivity of PR3 ANCA for active Wegener's is 91% and specificity 99%.⁵ However, a small number of patients with vasculitides such as microscopic polyangiitis and Churg–Strauss syndrome may test positive for PR3 ANCA.⁶ In addition, a positive PR3 ANCA may only be expressed late on in the disease. Hence, we suggest that in patients with active, severe disease who present to an emergency department, autoantibody testing should be used early, as it is more likely to show an abnormality. Biopsy can be performed once the patient is stable.

Fulminant disease may require dialysis, plasma exchange and, unlike other vasculitides, cyclophosphamide, making an accurate diagnosis important. Before its introduction, the median time from presentation to death was 5 months, typically from necrotising glomerulonephritis.⁷ This highlights

Classification criteria for Wegener's granulomatosis (sensitivity 88%, specificity 92%)

1. Development of oral ulcers, epistaxis or purulent nasal discharge
2. Chest radiograph showing nodules, fixed infiltrates or cavities
3. Urine analysis showing microscopic haematuria or red cell casts
4. Histological examination showing granulomatous inflammation in the wall of an artery or in the perivascular area (characteristically necrotising)

Abbreviations: PR3 ANCA, proteinase 3 antineutrophil cytoplasmic autoantibody

Figure 1 CT scan of the thorax of our patient showing a right upper lobe mass invading the chest wall with bilateral pulmonary nodules and an ipsilateral pleural effusion.

the need to perform urine analysis in patients with unusual ENT symptoms.

CONCLUSION

We remind emergency physicians to consider Wegener's granulomatosis in patients with ENT symptoms. It is important

to perform a full ENT examination, urine analysis and, if suspicious, a chest radiograph. It may be helpful to send a PR3 ANCA early in those presenting with active severe disease to reach a rapid diagnosis.

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