Case report:
A 23 year old female presented to the Emergency Department with a week long history of increasing shortness of breath, cough, fever and infrequent bouts of haemoptysis. She was normally fit and well and on no regular medications. On examination she had bilaterally reduced air entry with no audible crackles or wheeze.

Bloods:
- WCC 11.6
- Hb 84
- Platelets 601
- U+E normal
- INR 1.2
- GGT 78
- CRP 295

ABG:
- PH 7.37
- pO2 7.9
- pCO2 5.6
- HCO3 24
- BE -1.2

Observations:
- RR 22
- Sats 92% (15L NRB)
- Temp 37.8
- BP 110/68
- HR 96

She was initially treated for atypical pneumonia and commenced on IV fluids and antibiotics. She clinically deteriorated and was transferred to the Intensive Care Unit (ICU) where she was intubated and ventilated. A provisional diagnosis of Wegener's granulomatosis (WG) was made and prednisolone commenced. Her oxygen saturations remained poor and she was transferred for extracorporeal membrane oxygenation. Following bronchoscopy and biopsy she underwent a plasma exchange and was commenced on cyclophosphamide in addition to the steroids. She returned to our ICU and was successfully weaned from the ventilator and stepped down to the ward. She was subsequently discharged.

Background: WG - now known as granulomatosis with polyangiitis - is part of a large group of vasculitic syndromes. It is an autoimmune condition with anti-neutrophil cytoplasmic antibodies (ANCA) against small vessels.

Signs and symptoms:
- Lungs: Haemorrhage, Infiltrates, Nodules
- Joints: Arthralgia, Arthritis
- Skin: Pupura, Livedo-recticularis
- ENT: Epistaxis, Saddle nose, Deafness
- Kidneys: Glomerulonephritis

Investigations:
- CRP and ESR - typically high
- FBC – eosinophilia, anaemia
- Autoantibodies including ANCA if high clinical suspicion. A negative ANCA does not exclude vasculitis. The presence of PR3 ANCA is strongly suggestive of WG.
- Urinalysis: red cell casts
- Tissue biopsy: granulomatous inflammation
- Further imaging: chest CT or MR
- Assessment of critical organ function

Diagnosis: The American College of Rheumatology’s criteria – a patient must have 2 or more of:
- Nasal or oral inflammation
- Abnormal chest radiograph
- Urinary sediment
- Granulomatous inflammation on biopsy

Treatment: The natural history of untreated disease is rapidly progressive and usually fatal. The combination therapy of cyclophosphamide and prednisolone has resulted in a significant improvement in mortality and is effective in inducing remission\(^3\). Side effects of long term cyclophosphamide are extensive and include neutropenia, and consequently recent evidence suggests that following 3-6 months, azothiaprine or methotrexate can be used as a maintenance agent\(^4\). In cases of severe renal vasculitis or significant pulmonary haemorrhage, as in this case, plasma exchange may be used.

Conclusion: WG can present with vague symptoms, and this case highlights the need for timely diagnosis and early immunosuppression therapy, as the disease can result in significant morbidity and mortality.

References: