Hemoptysis with lung cavity — triple whammy

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A 37-year male presented with cough and dyspnoea for 9 months that had worsened over the past 7 days and was complicated by haemoptysis. Haemoptysis was moderate in amount with 3–4 episodes per day consisting of fresh red blood and was not associated with bleeding from any other site. There was no history of fever or weight loss. Dyspnoea was persistent and progressive to dyspnoea at rest on presentation. Two years ago, the patient had undergone 6 months of anti-tubercular therapy for sputum positive pulmonary tuberculosis.

On examination, the patient was conscious and oriented. His blood pressure was 110/70 mm Hg, pulse was 120/minute and respiratory rate was 34/min. Pallor was present but without any icterus, pedal oedema or lymphadenopathy. Respiratory system examination revealed vesicular breath sounds with bilateral crackles.

Investigations revealed Haemoglobin 9 gram% with normal organ function tests and hypoxemic respiratory failure. Chest X-ray and a CT examination of the chest are provided below (Figure 1 A–E).

Flexible bronchoscopy was essentially normal. Sequential bronchoalveolar lavage (BAL) fluid instillation became progressively bloodier upon aspiration and contained hemosiderin-laden macrophages confirming diffuse alveolar haemorrhage (DAH). BAL was negative for acid-fast bacilli (AFB), pneumocystis pneumonia (PCP), fungal mount, malignant cytology, and pyogenic/mycobacterial culture.

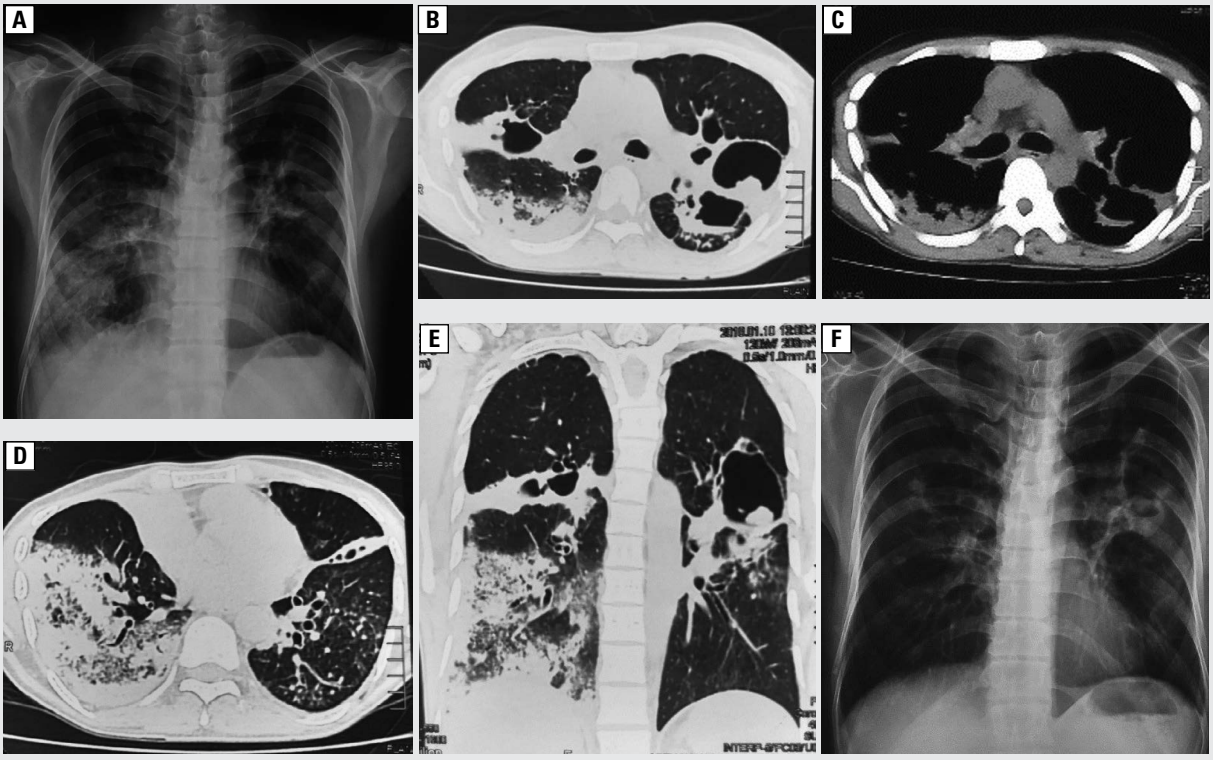

Figure 1. A. Chest X-ray showing bilateral cavitary lesions with right lower zone opacities; **B–E.** CECT chest suggestive of bilateral cavities with a fungal ball in the left upper lobe cavity. Also, bilateral dense alveolar opacities are present and are more prevalent in the right lower lobe; **F.** Chest X-ray showing resolution

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Serum precipitins and IgG against aspergillus were positive. Further probing revealed a history of intermittent episodes of epistaxis for 4 years which had spontaneously resolved. Further laboratory investigations revealed a positive c-ANCA by ELISA and anti-PR3 antibodies. Urine investigations were suggestive of 10–12 RBC with 24-hour protein of 400 mg%.

The American College of Rheumatology (ACR) 1990 classification criteria [1] for GPA was fulfilled by three out of four criteria (bloody nasal discharge, lung cavity, and ≥ 5 RBC in urine analysis). The 2017 proposed ACR/EULAR criteria require 5 points for a diagnosis; our patient satisfied 10 points (5 points for ANCA, 2 for lung cavity and 3 for epistaxis). This criterion is, however, suggested for clinical trials and the ACR suggests not to use it for diagnosis. Diffuse alveolar haemorrhage (DAH) presents with haemoptysis, anaemia or falling haematocrit, diffuse lung infiltration, and acute respiratory failure [2]. Our patient had all these features of DAH. It is difficult to diagnose DAH early because of its abrupt onset and rapid progression as well as because of its non-specific clinical symptoms and radiographic findings. The alveolar infiltrates can be unilateral, and a drop in hematocrit can be difficult to document [3].

The patient needed steroids and immunosuppressants, but also had CPA. The diagnosis of CPA requires consistent appearance in thoracic imaging (preferably by CT), direct evidence of Aspergillus infection or an immunological response to Aspergillus spp, and exclusion of some alternative diagnoses. In addition, at least 3 months of duration of disease are needed, even if inferred from or based on symptoms, or visualized on progressive radiological abnormalities [4]. Our patient had a lung cavity with positive serology to Aspergillus. Thus, the patient had a diagnosis of old treated pulmonary tuberculosis with GPA presenting as DAH with CPA. A cut-off of 10 mg prednisolone daily (or its equivalent) is arbitrarily considered to cause immunosuppression during clinical management. Intermittent higher levels of immunosuppression may accelerate progression of CPA if not controlled with antifungal therapy [4].

A case series found that seven ANCA-associated vasculitis patients developed invasive pulmonary aspergillosis when treated with steroids and immunosuppressants [5]. Therefore, they recommend that prophylactic antifungals should be given to high risk patients, especially those who are on immunosuppression.

The patient was given parenteral cyclophosphamide 750 mg for 1 day and methylprednisolone 1 g for 3 days under cover of intravenous voriconazole 200 mg every 12 hours.

Haemoptysis of the patient resolved in 2 to 3 days and room air saturation became 99%. The patient was discharged on oral prednisolone 1 mg/kg and oral voriconazole 200 mg twice daily and asked to follow up for subsequent pulses with cyclophosphamide. Chest X-ray at discharge (Figure 1F) showed resolution.

To conclude, cavitating lung diseases have varied etiologies and more than one apparent cause may be present simultaneously. Early diagnosis and treatment of diffuse alveolar hemorrhage can reduce mortality. Prophylactic antifungal treatment may be warranted in patients with an Aspergillosis infection who are put on immunosuppressant therapy.

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