Unusual association of diseases/symptoms

Organising pneumonia presenting as acute life threatening pulmonary haemorrhage

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Organising pneumonia, previously called bronchiolitis obliterans organising pneumonia is a clinicopathological entity of unknown aetiology, which has been reported with increasing frequency. Various modes of presentation have been described such as cough, fever, weight loss and alveolar opacities on chest radiograph. Haemoptysis as primary presenting symptom has only rarely been reported. The authors report a case in which massive life-threatening haemoptysis was the major presenting symptom. No aetiology was identified for the haemoptysis and the diagnosis was confirmed on postmortem histology. This case highlights the importance of considering organising pneumonia in the differential diagnosis of acute severe haemoptysis.

BACKGROUND

Organising pneumonia (OP) is a clinicopathological entity of unknown aetiology characterised histologically by granulation tissue plugs within the lumens of small airways extending into the alveolar ducts and airways.¹ OP has been reported with increasing frequency since the 1980s, both in an idiopathic form and in association with a variety of systemic or predisposing conditions. The typical patient presents with dyspnoea, cough, fever, weight loss and a chest radiograph with single or multiple alveolar opacities for which an infectious or neoplastic aetiology cannot be found. Haemoptysis as primary presenting symptom of OP has only rarely been reported.² We report a case of OP in which massive haemoptysis was the major presenting symptom.

CASE PRESENTATION

A 75-year-old male, smoker who was previously fit and well, presented with acute onset of episodes of massive haemoptysis amounting to a total of about 750 mls of clotted, dark blood mixed with sputum warranting blood transfusion. He denied long standing cough, dyspnoea, fever, weight loss or other systemic symptoms. There was no history of inhalation of toxic fumes or any drug allergies and he was not on any medications prior to presentation. There was no significant medical history apart from partial gastrectomy and duodenectomy for duodenal ulcer several decades back. At presentation, he was tachypnoeic, hypoxic, tachycardic, afebrile and chest examination was unremarkable.

He was admitted to medical ward where he became increasingly hypoxic and within few hours went into respiratory arrest following another bout of haemoptysis. Resuscitation team was alerted and he was intubated on the ward and taken to intensive care unit and ventilated but continued to have blood stained tracheal aspirate. His chest examination revealed bilateral crackles predominantly on left side.

INVESTIGATIONS

Chest radiograph showed patchy consolidation of the right lower zone and air space shadowing of left lower zone (figure 1a). Laboratory data on admission showed slightly elevated C reactive protein (15 mg/dl), low haemoglobin, normal coagulation, normal liver and kidney function and a positive troponin I (0.36). Repeat chest radiograph showed more diffuse non-homogenous air space opacification (figure 1b). CT thorax with contrast (figure 1c) showed extensive consolidation in the left lower lobe presumably secondary to haemorrhage (figure 1d–f), no active site of bronchial artery bleeding was demonstrated. There was fluid/debris in the major airways.

Electrocardiogram was normal sinus rhythm and echocardiogram was normal. Fibreoptic bronchoscopy showed fibrinous blood clots in the airways. Bronchoalveolar lavage (BAL) demonstrated reactive bronchial epithelial cells and macrophages. BAL was negative for acid and alcohol fast bacilli and one sample showed scanty growth of *Candida*. Bronchial biopsy was obtained but the sample was insufficient. A rigid bronchoscopy was also done which helped in evacuating some of the blood clots in the airways thereby improving his ventilation. Subsequently follow-up flexible bronchoscopies were done frequently which revealed further clots in the airways but no evidence of inflamed bronchioles. In the due course serial blood tests revealed raising inflammatory markers, low complement levels (C3-0.66 g/l, C4-0.11 g/l), mildly abnormal liver function and raised blood urea with normal creatinine. However his antinuclear antibody, extractable nuclear antigens, antineutrophilic cytoplasmic antibody and antiglomerular basement membrane antibody screen were negative. Serology for *Mycoplasma* was negative. Urine analysis showed no haematuria or proteinuria. One central venous line tip culture grew insignificant coagulase-negative *Staphylococcus* but other blood cultures were negative.

Figure 1 (a) Chest x-ray on admission. (b) Chest x-ray 1 week later. (c) (Blood clot/debris in airways). (d) Bilateral (mainly left lung) changes presumably due to haemorrhage. (e) Consolidation presumably due to haemorrhage. (f).

TREATMENT

Patient was treated with broad-spectrum antibiotic (piperacillin-tazobactam) and intravenous hydrocortisone at a dose of 300 mg daily for 3 days, which was reduced to 150 mg and given for a total duration of 1 week. He was intubated, ventilated and managed in the intensive care unit. A course of intravenous methylprednisolone was started on 14th day of hospital admission considering the possibility of OP, following negative results from BAL and vasculitis screen.

OUTCOME AND FOLLOW-UP

He was increasingly hypoxic requiring high-pressure ventilatory support and therefore the decision was to put a ceiling to ventilation. A possibility of hypoxic brain damage secondary to respiratory arrest was suspected because he remained unresponsive when off sedation. CT scan of brain showed evidence of age related cerebral atrophy. However, in view of failure to improve in neurological

status, a consensus opinion from neurologist and critical care physicians was that of a hypoxic brain damage. His condition worsened progressively and in view of poor prognosis a decision was made to withdraw the treatment and he died on the 19th day of admission. A Coroner's postmortem was requested as no established cause was found to explain the significant haemoptysis.

Postmortem findings

There were bilateral pleural effusions and there were propagating thromboemboli confined only to the lower lobe of the right lung. The cut surface of both the lungs showed fibrosis with extensive pulmonary oedema notably in the upper lobes without any consolidation. Histology confirmed the presence of pulmonary thromboembolism (figure 2) mostly confined to the larger blood vessels, which are recent in duration. Focal evidence of some organisation noted occasionally in the thrombi. There was no histological evidence of either haemorrhagic

Figure 2 Pulmonary thromboembolus.

Figure 3 Organising myxoid fibrinous plugs in intra-alveolar spaces (Masson's bodies) (x10).

infarcts or vasculitic pathology such as Wegener's granulomatosis. The other significant finding in both the lungs was the presence of organising exudates in the form of myxoid fibrous connective tissue within the alveolar air spaces (Masson's bodies) (figures 3 and 4). There was also a degree of thickening of the alveolar septae with

no histological evidence consistent with interstitial lung disease pathology. There were foci of diffuse alveolar damage with formation of hyalin membrane in the histological block sampled from the lower lobe of the left lung. Cytochemical stains (martius/scarlet/blue and Van Geison) (figure 5) showed the presence of organising

Figure 4 Masson's bodies (x40).

Figure 5 Elastic Vangeison stain (x40).

fibrous connective tissue with elastic damage of the alveolar walls confirming the presence of Masson's bodies. There was no significant fibrin demonstrated by the cytochemical stains. There was no histological evidence of consolidation. No neoplastic masses were identified. The histological features were in keeping with OP with pulmonary thromboemboli formation.

DISCUSSION

Lange initially described OP in 1901. Epler *et al*¹ published the largest case series of OP in 1985 and since then numerous reports have been published elucidating the various clinical presentations of this condition. The illness occurs in both men and women (aged 20–70 years) and is not known to be related to smoking.³ The typical

presentation is of a subacute or a chronic clinical course with fever, cough, weight loss, dyspnoea and less commonly chest pain and haemoptysis.^{4 5} The medical literature describes scanty haemoptysis⁴ in OP rarely, but to our knowledge there are only few case reports of this condition presenting with large quantity of haemoptysis.² We report this case of what we believe to be the first case of OP presenting with acute life threatening pulmonary haemorrhage. OP is defined⁶ pathologically by presence of granulation tissue composed mainly of fibroblasts and connective tissue matrix within the lumen of distal air spaces (bronchioles, alveolar ducts and alveoli). OP may be idiopathic (which is associated with a better prognosis)⁵ or secondary to bacterial (*Mycoplasma*) or viral infections (human immunodeficiency virus, herpes simplex virus), pharmacological agents (nitrofurantoin, sulfasalazine),⁷ chemotherapy, radiotherapy⁸ and connective tissue disorders.

Our patient was not on any drugs prior to his illness, there was no clinical history or findings to suggest any vasculitic process. Negative urine analysis, normal serum creatinine, normal initial inflammatory response, negative vasculitis screen and no vasculitic process on histological examination confirmed this. Normal inflammatory markers on admission, rapid onset of haemoptysis in the absence of systemic illness and failure to isolate any organism on repeated cultures makes infectious aetiology unlikely. The contrast CT thorax failed to identify the source of bleeding. This case highlights the importance of considering OP in the differential diagnosis of massive life-threatening haemoptysis in previously healthy individuals. Patients with OP have a >65% cure rate on corticosteroid therapy in most case series.^{1 9} Although, our patient was treated with steroids initially, because of the diagnostic uncertainty, an appropriate dose for treatment of OP was not given timely. An open lung biopsy would have been useful in our case as in some case series, its risk has been justified given the benefits of timely steroid therapy.^{10–12}

Learning points

- ▶ OP should be considered in the differential diagnosis of unexplained massive life-threatening haemoptysis in previously healthy individuals especially if other cause is not identified.
- ▶ OP can present acutely and deteriorate rapidly.
- ▶ High index of suspicion is important to make a diagnosis so that early therapy can be instituted.

Competing interests None.

Patient consent Obtained.

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