CASE REPORT

Unexpected case of pneumomediastinum and subcutaneous emphysema: primary or secondary aetiology?

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SUMMARY

A 77-year-old man was admitted with a relapse of antineutrophil cytoplasmic antibody-positive vasculitis with pulmonary involvement and acute kidney injury. There was a background of pulmonary fibrosis (non-specific interstitial pneumonia type pattern) and superadded pulmonary haemorrhage, acute pulmonary oedema and sepsis. The patient was intubated for 4 days and remained dependent on high flow oxygen and continuous positive airway pressure after extubation. A chest radiograph performed 2 weeks after extubation demonstrated unexpected, extensive pneumomediastinum and subcutaneous emphysema. This was confirmed on CT which raised the possibility of a tracheal defect at the level of the prior endotracheal tube cuff position. Tracheal injury was considered clinically unlikely due to the considerable interval since extubation and a short, uneventful intubation period. The cardiothoracic team recommended a diagnostic bronchoscopy but this was felt too high risk by the clinical team. The cause of pneumomediastinum and subcutaneous emphysema remained indeterminate.

BACKGROUND

The advantages of a multidisciplinary approach to problem solving are demonstrated here with key decision-making elements brought together by respiratory, renal, radiology and critical care specialists.

Despite several investigations, it was difficult to ascertain whether the aetiology was primary (spontaneous), secondary with respect to predisposing lung conditions (interstitial lung fibrosis, infection) or secondary iatrogenic causes (positive airway pressure, tracheal injury from endotracheal tube placement). Determination of the exact aetiology did not ultimately impact on the clinical outcome. However, this case highlights the difficulties investigating such complex patients.

Imaging plays an increasingly important role in the management pathway of pneumomediastinum and subcutaneous emphysema but we present here a discussion of some of its uses and limitations.

CASE PRESENTATION

A 77-year-old man presented with a relapse of myeloperoxidase antineutrophil cytoplasmic antibody-positive vasculitis, which was first diagnosed within the preceding month.

The patient had been diagnosed with lung fibrosis 5 years ago after being referred to the interstitial lung disease service at a specialist chest hospital. He underwent video-assisted thoracoscopic surgery in order to obtain a biopsy for which the pathology showed mixed inflammation and fibrosis. This was compatible with the imaging features of multifocal ground glass changes and reactive mediastinal lymph node enlargement and overall in keeping with a fibrosing non-specific interstitial pneumonia (NSIP) pattern.

There was stable exercise tolerance documented for the preceding 3 years and no specific treatment for fibrosis was commenced until this admission, on which he was started on prednisolone. There was a medical history of chronic immune thrombocytopenia and idiopathic thrombocytopenic purpura for which he had received immunomodulatory drugs and steroids. The respiratory team determined that the immune suppressant therapy had most likely modified his presentation of lung fibrosis and was associated with his vasculitis. There was a long history of atrial fibrillation, which was being treated with warfarin.

The patient had never smoked and he worked in a car scrapyard all his life with chronic exposure to metal and paint dust but denied asbestos exposure. He kept pet chickens, ducks and cats.

His current symptoms were bilateral pitting oedema, shortness of breath and haemoptysis.

On examination, he was dyspnoeic with difficulty completing sentences and had a dysfunctional breathing pattern. There was no clubbing or skin rash. On auscultation of the chest there were widespread inspiratory crackles and harsh breath sounds.

INVESTIGATIONS

CT chest (day 5 of admission)

The patient was intubated from days 5–8 of admission. A CT performed on the day of intubation showed an NSIP fibrosing pattern with progressive multifocal ground glass changes and new consolidation. Superimposed features of pulmonary interstitial oedema were seen on a background of cardiomegaly and there were new pleural effusions. The overall picture concluded progressive pulmonary haemorrhage with fluid overload and superadded infection on a background of extensive fibrosis (video 1). The suture line from previous

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Video 1 Axial CT Thorax. Satisfactory position of the endotracheal tube terminating above the level of the carina. Peripheral reticular changes, bronchiectasis and extensive ground glass attenuation in keeping with fibrosis (non-specific interstitial pneumonia type pattern). Cardiomegaly, new pleural effusions and interlobular septal thickening consistent with pulmonary interstitial oedema and fluid overload. Progressive patchy areas of consolidation from superadded infection and pulmonary haemorrhage.

VATS-assisted biopsy is seen in the lingula of the left upper lobe ([figure 1](#)).

Bronchoalveolar lavage (day 6 of admission)

Enterobacter cloacae was cultured and results were negative for PCR.

CXR (day 8 of admission)

The patient underwent an uneventful extubation and no new findings were seen on the chest radiograph after removal of the endotracheal tube. The patient was subsequently dependent on high-flow nasal oxygen and helmet continuous positive airway pressure.

CXR (day 25 of admission)

After a relatively stable course, the patient developed a sudden deterioration in oxygen saturations to 85% and required 15 L of oxygen to maintain low saturations. He experienced shortness of breath but had no cough, haemoptysis or chest pain. New extensive subcutaneous emphysema and pneumomediastinum was seen on the chest radiograph which was an unexpected finding with striking appearances ([figure 2](#)). As there was no history of trauma and the cause was unclear, radiology was consulted and a CT was arranged to further investigate.

CT chest (day 25 of admission)

Extensive subcutaneous emphysema and pneumomediastinum was confirmed ([video 2](#)). A possible tracheal defect was seen at

Figure 1 Axial CT thorax. Lingula lobe suture from previous biopsy with video-assisted thoracoscopic surgery (VATS).

the level of the prior endotracheal cuff position ([figure 3](#)). There was no pneumothorax or evidence of oesophageal rupture. Pulmonary changes of haemorrhage and consolidation were stable and the pleural effusions had improved.

Cardiothoracic surgical team consulted

Diagnostic bronchoscopy was initially recommended but subsequently not performed as it was deemed too high a risk by the clinical team.

DIFFERENTIAL DIAGNOSIS

Tracheal injury was the initial concern in view of the CT findings but was considered clinically unlikely due to an uneventful,

Figure 2 Frontal chest radiograph. Satisfactory position of the right peripherally inserted central catheter terminating at the mid superior vena cava. New extensive gas in the cervical, axillary and chest wall soft tissues in keeping with subcutaneous emphysema. There is also pneumomediastinum with gas outlining the aortic knuckle.

Video 2 Axial CT Thorax. Extensive gas in the neck and chest soft tissues in keeping with subcutaneous emphysema. Florid pneumomediastinum but no pneumothorax. A possible linear defect is indicated by the pointer at the 5 o'clock position in the trachea but no gross defect is seen. The pleural effusions have decreased but there is ongoing widespread consolidation.

short intubation period and considerable interval of 2 weeks since extubation. This could not be confirmed either way on the basis of imaging.

A more likely iatrogenic cause included mechanical ventilation support and oxygen therapy, causing increased pressure gradients.

Figure 3 Axial CT thorax. Possible defect in the trachea coinciding with the level of the previous endotracheal balloon cuff.

Spontaneous pneumomediastinum and/or subcutaneous emphysema are considered where no secondary lung or iatrogenic causes are found. Underlying interstitial lung disease increases the likelihood of gas escaping into the soft tissues if there are weakened bronchoalveolar walls or ruptured bulla. Valsalva manoeuvre during coughing can also lead to the same phenomenon.

TREATMENT

The patient received methylprednisolone, plasma exchange (PLEX 5X) and intravenous cyclophosphamide and went on to have further plasma exchange and a further five cycles of cyclophosphamide during his admission. Prednisolone was started during this admission for his lung fibrosis having not been on specific treatment for this previously.

Due to the development of sepsis, meropenem and clarithromycin were administered.

Following the development of pneumomediastinum and subcutaneous emphysema, treatment was supportive with ventilator support not reinstated due to the possibility of an air leak.

OUTCOME AND FOLLOW-UP

The clinical outcome was felt to be determined by the progressive, treatment-refractory pulmonary fibrosis and the patient was referred to the community palliative care team.

Appointments were scheduled for the vasculitis and critical care outpatient clinics. The patient was stable on home oxygen initially but died 8 days following discharge from hospital.

DISCUSSION

Pneumomediastinum refers to the presence of gas within the mediastinal space. When it occurs spontaneously, it is known as Hamman's syndrome and was first described in 1939.¹ Hamman's sign refers to a phenomenon whereby crepitus is heard to coincide with the heartbeat on auscultation.²

Spontaneous pneumomediastinum occurs more commonly in young adult patients when exposed to sudden changes in pressure within the intrathoracic cavity.

Subcutaneous or surgical emphysema refers to the presence of gas within the subcutaneous tissues, typically arising in the supraclavicular/cervical soft tissues before spreading to other parts of the body. It is more commonly associated with trauma (blunt or penetrating) or surgery and may occur from iatrogenic causes such as tracheal or intercostal tube insertion. If it occurs spontaneously due to the rupture of a subpleural cavity or bulla, it is usually associated with a pneumothorax. However, subcutaneous emphysema in the absence of pneumothorax, as in our case, has only been rarely described.¹

Other causes of secondary pneumomediastinum include rupture of a hollow viscus, tissue dissection originating from pneumothorax and pulmonary or mediastinal gas-forming organisms.³ In respiratory pneumomediastinum, a phenomenon known as the Macklin effect is seen whereby alveolar gas centripetally dissects via the pulmonary interstitium towards the hila and mediastinum, leading to linear collections that can be seen on CT to run along the bronchovascular sheaths.⁴

The chronic lung conditions of this patient most likely predisposed him to gas escaping into anatomical spaces and soft tissues due to there being an increased stiffness of his lungs. However, this could not be proven and led to the uncertainty of whether this was spontaneous or secondary aetiology.

Subcutaneous emphysema and/or pneumomediastinum following intubation or extubation is a rare occurrence. Causes

include a tear resulting from passage of the endotracheal tube or from overinflation of the cuff.⁵ Variants in the trachea such as bullae, clefts or laryngocoeles can also serve as lead points for mucosal dehiscence secondary to damage from the tube.

In the case of our patient, a possible tracheal defect seen on CT coincided with the position of the previously sited cuff balloon. The length of time since extubation and any technical difficulties encountered with respect to intubation should be included in the details of the scan request in order for the reporting radiologist to formulate a differential where appropriate. Cross-sectional imaging lacks the spatial resolution to reliably diagnose an

intimal tear of the trachea, and so unless there is a gross defect, only the possibility of an intimal tear should be raised.

The role of imaging in this context was appropriate to investigate for evidence of pneumothorax, oesophageal rupture or tracheo-oesophageal fistula but tracheal tears can be confirmed only by direct visualisation at tracheobronchoscopy. The use of cross-sectional imaging is additionally suitable to look for haematoma, collections or evidence of infection and can delineate the spaces within which gas is present.

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Learning points

- Pneumomediastinum and subcutaneous emphysema have primary (spontaneous) and secondary causes.
- Pneumomediastinum and subcutaneous emphysema are rare complications following endotracheal tube placement or removal. These conditions may present with minimal symptoms and should be interpreted in the clinical context.
- Patients with chronic interstitial lung disease may develop pneumomediastinum more readily following rupture of bronchoalveolar walls or subpleural spaces during increased intrathoracic pressure, for example, coughing or mechanical ventilation.
- Tracheal tears should be investigated with tracheobronchoscopy where clinically appropriate as CT often lacks the spatial resolution to confirm an intimal or mucosal tear.
- The use of CT is appropriate to investigate for concurrent pneumothorax and can exclude alternative causes such as oesophageal rupture.

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