CASE REPORT

Pneumothorax necessitans in a patient with trapped lung and rheumatoid arthritis

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SUMMARY

The authors report the case of a patient with a background of trapped lung following thoracocentesis who developed an anterolateral intercostal pneumothorax resulting in a pneumothorax necessitans (PN). Our purpose is to highlight the pathophysiology and interesting radiological features associated with PN in trapped lung. Our case is particularly unique due to the subacute nature of its presentation in a patient with rheumatoid arthritis and unusual pre-existing lung pathology.

BACKGROUND

Trapped lung is an unusual and uncommon phenomenon seen as a consequence of pleuritic fibrosis or granulomatosis induced by a preceding inflammatory process leading to stiff pleura in a partially expanded lung. While single or multiple aetiologies may be implicated, the diagnosis of trapped lung should always be considered in those with a chronic and persistent benign pleural effusion. The condition lies at one end of a spectrum under the umbrella term of 'Unexpandable lung'.¹ This describes restriction of the visceral pleura resulting in atelectasis or lobar collapse. Figures regarding incidence and prevalence are unknown due to the paucity of reports in the literature, but this process often leads to distinct radiological features.

A more unusual phenomenon than unexpandable lung is the pathogenesis of pneumothorax necessitans (PN), which describes a pre-existing pneumothorax fistulating through the thoracic wall into a subcutaneous airpocket. The latter forms as a result of air escaping the described pneumothorax through a point of weakness in the thoracic wall leading to the formation of a subcutaneous 'bleb'.

Cases of PN within the literature have been described in the context of an empyema, secondary spontaneous pneumothorax and complication of closed tube thoracostomy.^{2–4} Both cases referred to the patients with systemic symptoms concordant with an intercurrent infective inflammatory process.

Our unique case describes a highly unusual dual presentation of PN with co-existing trapped lung on a background of pulmonary rheumatoid arthritis (RA). Our patient presented subacutely following the resolution of chronic benign pleural effusion. As far as the authors are aware, this is the first such documented case in the literature.

CASE PRESENTATION

A 65-year-old gentleman, a former marine engineer and longstanding smoker, with a background of

chronic RA and a persistent right-sided pleural effusion, attended a routine outpatient follow-up appointment for his RA at the Queens Hospital, Romford, UK, in July 2012.

Six months previously, he had an unsuccessful talc pleurodesis for recurrent right-sided pleural effusions, following which he had further chest drainage. The effusion resolved; however, the patient developed a small pneumothorax as a complication of the drain. In addition, the drain site had not healed completely and was leaking fluid both during drainage and for some days after the drain had been removed. As he was not in any respiratory compromise, he was observed for a few days along with regular dressing changes and was discharged with advice and reassurance. Within weeks he suffered a myocardial infarction and had a subsequent pneumonia while in hospital.

In July 2012, he had a routine follow-up appointment with his rheumatologist regarding his long-standing RA. It was here that the patient revealed that a subcutaneous reducible mass had gradually developed at the point of the healed drain site over a period of weeks to months, but did not present to a doctor with this complaint before. He denied of having any systemic symptoms. Subsequent chest radiograph and CT imaging were arranged.

INVESTIGATIONS

The patient was asymptomatic and not in any respiratory distress, his vital observations were normal. Chest x-ray (figure 1) revealed a right-sided pneumothorax with partial right lower lobe collapse

Figure 1 Chest x-ray.

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Figure 2 CT axial view.

Figure 4 CT sagittal view.

and with substantial resolution of the pleural effusion seen on a previous film. CT of the chest with contrast was performed, which revealed thickened pleura, loculated pneumothorax on the right side in communication with a subcutaneous air cavity 5 cm in diameter within the lateral chest wall (figures 2–4).

His blood tests revealed a normal full blood count, normal renal function, erythrocyte sedimentation rate of 9 and C-reactive protein of 18. His most recent pleural fluid analysis—which had been taken from the pleural space a few weeks before and now occupied by air—had shown no evidence of malignancy or infection and was exudative. Rheumatoid factor (RF) at the time of pleural fluid drainage was recorded as being >120 IU/l, and the white cell differential on full blood count was in the normal range.

DIFFERENTIAL DIAGNOSIS

The key differential in this case would be that of a traumatic pneumothorax, particularly following the drainage of an effusion and is known as post-thoracocentesis pneumothorax.¹ In

the case of this patient's trapped lung, it appears that while the effusion had partially resolved following previous drainage, there was a further phase of resolution, and despite the presence of a negative pressure pleural space, the cavity had been filled with air. This appears to have been caused by a spontaneous pressure-dependent fistula between the pleural space and the trapped lung, and is unlikely to be a case of lung trauma or air introduction, but rather a combination of unexpandable lung and formation of fistulous air tracts not requiring treatment.

Unlike a conventional pneumothorax, however, the trapped lung does not change in size during ventilation. Contrast this with the pneumothoracocoele, where the subcutaneous air pocket alters in size according to changes in intrathoracic pressure. The lung parenchymal border is also more defined due to the thickened visceral layer; although clearly seen on CT, it may also be visible on chest radiographs.

DISCUSSION

The umbrella term of unexpandable lung also describes 'lung entrapment', a related entity to trapped lung, which are both pathophysiologically similar, in that there is mechanical obstruction to lung expansion by restrictive visceral pleura. However, while the former is caused by postinflammatory fibrosis and collagen deposition, the latter is caused by active inflammatory pleuritis reducing expansibility. It is therefore the absence of active inflammation that differentiates trapped lung from lung entrapment. This differentiation can also be observed on the histological scale with an increase in collagen formation in trapped lung, as the inflammatory process is substituted by fibrosis.¹

The causes of trapped lung are manifold and include post coronary artery bypass grafting, rheumatoid pleuritis, tuberculosis, uraemia, radiation and prior empyema. Pleural fluid in a trapped lung tends to be a transudate with a decrease in both lactate dehydrogenase and protein levels. The persistent effusion appears to be caused by the constant negative pressure gradient existing between the pleural space and the neighbouring tissues.¹

X-ray and CT are the key imaging modalities used in the diagnosis. Chest radiographs in trapped lung tend to show large volume pleural effusions or pneumothoraces that may have the potential to cause alarm in the emergency department, even

Figure 3 CT coronal view.

Figure 5 Pathogenesis of pneumothorax necessitans.

though the patient may be asymptomatic. CT imaging would reveal a markedly thickened visceral pleural ‘peel’. The identification of the visceral pleura and its thickening accompanied by lobar collapse or atelectasis would be diagnostic of an unexpandable lung. Several studies contribute to the evidence that video-assisted thoracoscopic surgery is the preferred method in the treatment of trapped lung.^{5–8}

Due to the paucity of literature and low incidence of PN, there is little information on the pathogenesis of a subcutaneous airpocket in communication with an underlying pneumothorax. In the case of trapped lung, PN can only develop after the causative effusion has resolved and the fluid has been substituted by air. How air enters this space to start with is not fully certain. The most plausible theory is that a bronchopleural fistula formed over the course of a chronic effusion allows the introduction of intrapulmonary air to accumulate in the negative pressure cavity formed by the trapped lung.³ The formation of a subsequent pneumothoracoele occurs due to a defect in the chest wall, for example, following chest drainage, allowing an extrathoracic abscess to be formed. It is this structure that becomes air filled when the empyema resolves in the presence of a trapped lung. In our patient, this weakness in the chest wall came about at the point of previous chest drainage. Figure 5 outlines what the authors believe to be the key stages in the development of PN.

The interesting diagnostic conundrum in the case of our patient is whether trapped lung was initiated by an initial infective process or a rheumatoid-mediated inflammation or indeed a combination of both. The chronic and recurrent effusions suffered by our patient with high titres of RF may have triggered pleural fibrosis.⁹ On the other hand, pneumonia may in fact have been the one and only aetiology leading to pleural fibrosis. With regard to the formation of PN, a plausible theory would be that the pneumonia suffered by our patient along with coughing and therefore increased intrathoracic pressures

generated from this may have triggered the formation of a bronchopleural fistula and the subsequent PN.

Treatment of PN in the context of a trapped lung is therapeutically challenging. There is no consensus on the reduction of a pneumothoracoele in cases of PN. In our patient, cardiothoracic intervention has not been offered, but instead conservative observation and symptomatic treatment was suggested.

Learning points

- ▶ Pneumothorax necessitans (PN) occurs due to an abnormal communication between a pre-existing pneumothorax and subcutaneous air collection.
- ▶ Radiological features of trapped lung are distinct with the presence of a thickened fibrous peel outlining the lung.
- ▶ PN can cause alarm to both the patient and the clinician on the basis of its appearance and should always be investigated following a thorough history.

Competing interests None.

Patient consent Obtained.

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