MMJ

CASE REPORT

Spontaneous pneumomediastinum in idiopathic pulmonary fibrosis

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Spontaneous pneumomediastinum is an uncommon occurrence and is usually self-limiting. We present an 84-year-old gentleman with idiopathic pulmonary fibrosis (IPF) presented with worsening breathlessness and a worsening dry cough with no history of haemoptysis. A computerised tomography (CT) scan of the thorax revealed marked worsening of the pulmonary fibrotic changes as well as extensive pneumomediastinum extending throughout the length the mediastinum. A diagnosis of spontaneous pneumomediastium was made. The patient was managed conservatively with oxygen and was discharged home after his dyspnoea gradually improved.

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Background

Spontaneous pneumomediastinum is an uncommon occurrence and is usually self-limiting. Spontaneous pneumomediastinum is the presence of free air around mediastinal structures and is thought to be secondary to alveolar or honeycomb cyst rupture due to raised intrapulmonary pressure on coughing, blunt force trauma to the chest, asthma, various types of interstitial lung diseases, including interstitial pneumonia associated with connective tissue diseases, including dermatomyositis and rheumatoid arthritis.[1]

We herein report a rare case of spontaneous pneumomediastinum with thoracic-wall subcutaneous emphysema occurring in an elderly gentleman with idiopathic pulmonary fibrosis (IPF).

Case report

An 84-year-old gentleman with a background of IPF on long-term oxygen therapy, presented to a scheduled outpatient appointment complaining of worsening dyspnoea and weight loss. The patient described recent worsening shortness of breath after a persistent coughing bout. He was unable to perform his activities of daily living unassisted and was house bound in view of the progressive dyspnoea (Modified Medical Research Council Dyspnoea scale 4). There was no history of haemoptysis, pleurisy, chills, rigors or fever suggesting an underlying infective pathology. Medical history was otherwise significant for glaucoma and hypothyroidism on replacement therapy. Two years prior, he was found to be hypoxaemic on ambulation and thus was initiated on 2 L/minute home oxygen therapy which he was compliant to.

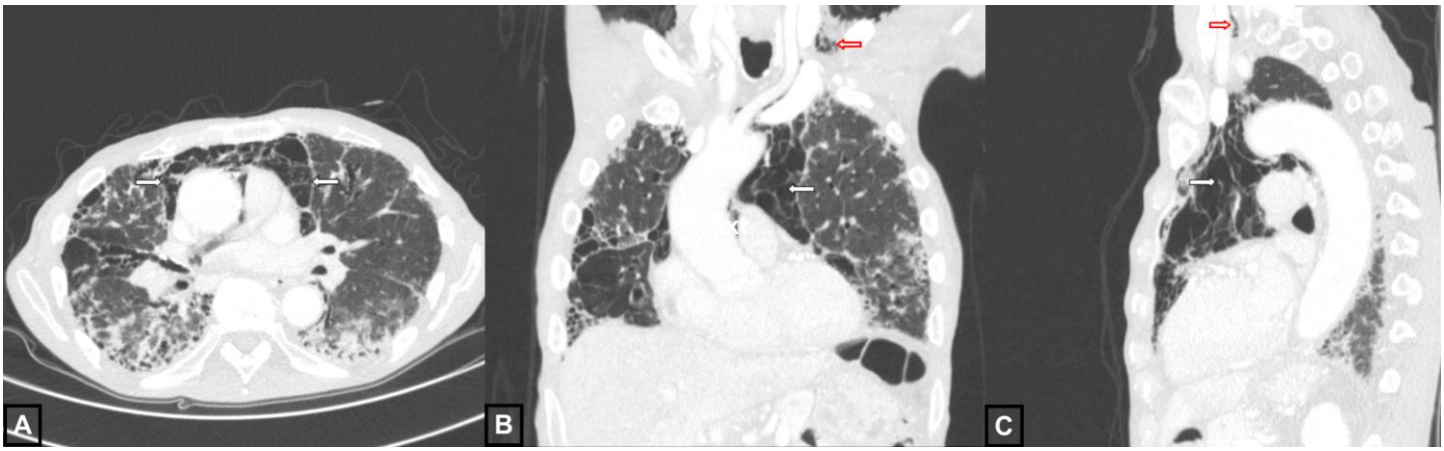
The patient was an ex-smoker, having stopped smoking 6 years ago, with a 35-pack-year history. On clinical examination he was clubbed, cachectic and breathless, but was not in respiratory distress. He was otherwise haemodynamically stable with oxygen saturations of 87% on room air and diffuse fibrotic crepitations in both lung fields. The patient was unable to withstand repeat pulmonary function testing on the day of presentation.

An urgent computerised tomography (CT) scan of the thorax revealed marked worsening of the pulmonary fibrotic changes especially in the lung bases with honeycombing, traction bronchiectasis and diffuse interstitial thickening. It also showed extensive pneumomediastinum (white arrows, figure a-c) with air extending throughout the length of the mediastinum and into the subcutaneous tissues of the neck along the vascular sheath (red arrows, figure b-c). In retrospect, there were no clinical signs of surgical emphysema of the anterior chest wall and neck and Hamman's sign was not appreciated.

On further questioning, the patient had not undergone any recent tracheobronchial or gastroscopy procedures effectively ruling out oesophageal rupture in absence of features of shock. There was no evidence to suggest pulmonary or mediastinal infection from gas-forming organisms or recent trauma to chest. Thus, a diagnosis of spontaneous pneumomediastium was made and it was managed conservatively with oxygen and bed rest. The patient was reviewed at outpatients after a few days with radiological improvement.

Figure 1 **Spontaenous pneumomediastinum in a patient with idiopathic pulmonary fibrosis.**

(a) Axial, (b) coronal, (c) transverse CT images shows extensive pneumomediastinum (white arrows) and subcutaneous emphysema (red arrows) without any co-existing pneumothorax.



Discussion

Spontaneous pneumomediastinum is an uncommon occurrence and patients usually present with pleuritic chest discomfort with worsening dyspnoea, odynophagia and neck pain. Subcutaneous emphysema is commonly noted, particularly in the neck. Hamman's sign, a crunching sound, synchronous with the patient's heartbeat, could be noted on examination.

Air tends to track within the mediastinal planes, ranging from the submadibular space, retropharyngeal space and the vascular sheath in the neck to the retroperitoneal space and pelvis. Thus, pneumomediastinum is frequently associated with other forms of extra-alveolar air, such as pneumopericardium, pneumothorax, subcutaneous emphysema, pneumoretroperitoneum and pneumoperitoneum as a result of these communications.[2]

CT imaging is the diagnostic gold standard in suspected spontaneous pneumomediastinum.[3] Spirometry and peak expiratory flow rate is contraindicated in patients as it may exacerbate spontaneous pneumomediastinum in view of intrathoracic pressures.[4] Uncomplicated spontaneous pneumomediastinum is managed conservatively with bed rest, adequate analgesia and avoidance of manoeuvres than increase intrathoracic pressures such as Valsalva manoeuvre.[5] In patients who are increasingly symptomatic, high-flow oxygen inhalation therapy is used in order to enhance its re-absorption by nitrogen washout.[6]

References

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