Spontaneous pneumothorax: epidemiology, pathophysiology and cause

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ABSTRACT: Spontaneous pneumothorax represents a common clinical problem. An overview of relevant and updated information on epidemiology, pathophysiology and cause(s) of spontaneous (primary and secondary) pneumothorax is described.

KEYWORDS: Epidemiology, pathogenesis, pneumothorax

Primary spontaneous pneumothorax (PSP) has an incidence of 7.4 to 18 cases (age-adjusted incidence) per 100,000 population each year in males, and 1.2 to 6 cases per 100,000 population each year in females [4, 5]. PSP typically occurs in tall, thin subjects. Other risk factors are male sex and cigarette smoking. Contrary to popular belief, PSP typically occurs at rest; avoiding exercise, therefore, should not be recommended to prevent recurrences [6]. Precipitating factors may be atmospheric pressure changes (which may account for the often observed clustering of PSP) and exposure to loud music [8]. Almost all patients with PSP report a sudden ipsilateral chest pain, which usually spontaneously resolves within 24 h [2]. Dyspnoea may be present but is usually mild. Physical examination can be normal in small pneumothoraces. In larger pneumothoraces, breath sounds and tactile fremitus are typically decreased or absent, and percussion is hyper-resonant. Rapidly evolving hypotension, tachypnea, tachycardia and cyanosis should raise the suspicion of tension pneumothorax, which is, however, extremely rare in PSP.

Diagnosis can be confirmed in the majority of cases on an upright posteroanterior (PA) chest radiograph, which also allows an estimation of the pneumothorax size with good accuracy [9]. In cases with a small PSP, computed tomography (CT) may be necessary to diagnose the presence of pleural air. Routine expiratory chest radiographs are useless [10]. It is important to realise that a contralateral shift of the trachea and mediastinum is a completely normal phenomenon in spontaneous pneumothorax, and not at all suggestive for tension pneumothorax; this observation should therefore in no way influence treatment strategies [1].

Pathogenesis

The exact pathogenesis of the spontaneous occurrence of a communication between the alveolar spaces and the pleura remains unknown. Most authors believe that spontaneous rupture of a subpleural bleb, or of a bulla, is always the cause of PSP [11], but alternative explanations are available [12, 13]. Although the majority of PSP patients, including children [14], present blebs or bullae [15–18], it is unclear how often these lesions actually are the site of air leakage [19–21]. Only a minority of blebs are actually ruptured at the time
SECONDARY SPONTANEOUS PNEUMOTHORAX

A multitude of respiratory disorders have been described as a cause of spontaneous pneumothorax. The most frequent underlying disorders are chronic obstructive pulmonary disease with emphysema, cystic fibrosis, tuberculosis, lung cancer and HIV-associated Pneumocystis carinii pneumonia, followed by more rare but “typical” disorders, such as lymphangioleiomyomatosis and histiocytosis X (table 2). Because lung function in these patients is already compromised, secondary spontaneous pneumothorax (SSP) often presents as a potentially life-threatening disease, requiring immediate action, in contrast with PSP, which is more of a nuisance than a dangerous condition. The general incidence of SSP remains unresolved [35, 36].

These lesions may, therefore, predispose to PSP when combined with (largely unknown) precipitating factors; blebs and bullae indeed also occur in up to 15% of normal subjects [15–17]. New techniques such as fluorescein-enhanced autofluorescence thoracoscopy [37] or infrared thoracoscopy [38] may shed more light on this issue, and may be helpful in the detection of the culprit areas during thoracoscopy or surgery. It should be clear, however, that every therapeutic intervention with the purpose to prevent recurrences of PSP should include a pleurodesis technique, with or without an intervention at the level of the lung parenchyma [39].

resulting in acute respiratory failure, can also be present. Diagnosis is confirmed on a PA chest radiograph; in bullous emphysema, the differential diagnosis with a giant bulla can be difficult, necessitating CT confirmation [40]. As in PSP, air may enter the pleural space through various mechanisms: direct alveolar rupture (as in emphysema or necrotic pneumonia), via the lung interstitium, or backwards via the bronchovascular bundle and mediastinal pleura (pleuromediastinum). Recurrence rates usually are higher when compared to those for PSP, ranging up to 80% of cases, as is observed in cystic fibrosis [41].

STATEMENT OF INTEREST

None declared.

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