Primary Spontaneous Pneumothorax: An Update

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Primary spontaneous pneumothorax (PSP) is defined as a pneumothorax occurring spontaneously in a person without (known) underlying lung disease. PSP is common in young, tall, thin people without clinically apparent underlying lung disease. The incidence is estimated at 18 - 28 per 100,000 for males and 1.2 - 6 per 100,000 for females[1]. The cause of PSP is unknown, although it is mostly attributed to the rupture of a subpleural bleb or bulla[2]. The etiology of bulla and bleb formation is obscure. Subpleural blebs or bullae, which are designated as emphysema-like changes (ELCs), are seen in 75-100% of patients with PSP even in non-smoking PSP patients[3]. Macroscopic findings in the PSP group, according to the classification of Vandeschueren are: type I (normal findings); type II (pleuropulmonary adhesions); type III (bullae/blebs less than 2 cm diameter); and type IV (bullae >2 cm diameter) cases[3]. In a study of 94 patients treated with video-assisted thoroscopic surgery (VATS), 67 patients (71%) had a clear bullae in type III and IV. In 15 (16%) cases, (type II) pleuropulmonary adhesions were identified and in only 12 (13%) patients (type I) did thoracoscopy fail to reveal any abnormality[4]. With the development of VATS, blebs and bulla during thoracoscopy were detected in more than 76% of patients[3].

The actual site of air leakage, however, can be located at the ELCs which may be ruptured in some cases or elsewhere at the lung surface (“Pleural Porosity”). True visible air leaks at the site of the ELCs have been observed in a highly variable percentage of PSP patients undergoing thoracoscopy or thoracotomy[5]. Ayed et al[4] observed 94 patients with PSP who underwent VATS; leaking or ruptured blebs were seen in 24 patients (26%) at thoracoscopy. Light microscopy studies have shown that only 25% of the ELCs in PSP are truly ruptured, whereas in rest of the cases other lesions were present, referred to as ‘Pleural Porosity’[6, 8]. This porosity consists of mesothelial cell proliferation disrupure and elastofibrosis. Light microscopy has shown the actual site of air leakage at the site of ELCs in 15 patients (16%) and elsewhere on the lung surface in five other patients (5%)[4].

A variety of pathologic changes were seen at the lung apices. These changes include ELCs, airway inflammation, and emphysema. In a recent study, 74 out of 94 patients (79%) who required surgical intervention for persistent or recurrent PSP had a diagnosis of ELCs. Irregular emphysema, which was the most common type of emphysema identified, was seen in 14 patients[4]. The same study showed a pleural inflammatory reaction in patients with PSP, which is characterized by increases in parietal pleural tissue eosinophils and neutrophils and associated with pleural fibrosis. Another interesting observation in patients with PSP is that all patients had microscopic evidence of underlying lung disease in the excised apex of the lung. These results suggest that these lesions are the result of a degenerative process in the lung[4,7]. It seems important to care for the apex of the lung because spontaneous pneumothorax originates from dystrophic areas located in the apices of the lung, and not from any kind of pleural disease. These observations support the presence of underlying lung disease in the etiology of PSP. In addition, the inflammatory changes in the distal airways of smokers suggest that there is some degree of endobronchial obstruction involved in the pathogenesis of PSP. Endobronchial obstruction due to accumulation of inflammatory cells between the pulmonary parenchyma and the bronchial tree can induce overpressure in alveolar tissue which can lead to rupture of pulmonary parenchyma[9]. Light and electron microscopy of tissue obtained during surgery for PSP have revealed obstruction and stenosis of the distal airways due to bronchial wall inflammation and peribronchial fibrosis[3,4]. These findings suggest an obstruction check-valve mechanism, with air trapped in small airways because of the narrowed inflamed small airways, as the cause of PSP. Autofluorescence.
thoracoscopy allowed visualization of extensive lung areas with subpleural fluoresceine accumulation suggesting the presence of substantial areas of lung parenchymal abnormality[8].

The therapeutic challenge in the management of PSP is to prevent recurrence because understanding the exact pathophysiology of PSP in an individual is important. The recurrence prevention technique may differ accordingly. The recurrence technique focuses on the treatment of lung abnormalities such as ELCs or treatment of pleura. In type III and IV cases, VATS blebectomy/bullectomy plus apical pleurectomy or pleurodesis is effective[4,8]. Persistent air leak and recurrence rate of PSP were higher in type I cases after the excision of the apex and apical pleurectomy. This may indicate that the lung apices are not the actual site of the air leak; or air leakage occurs elsewhere at the visceral pleura. Therefore, simple apical excision and apical pleurectomy in these cases are not sufficient and perhaps additional talc poudrage to induce more pleural symphysis might be indicated. Other maneuvers to create pleurodesis and prevent recurrences include parietal pleural abrasion with dry gauze or any other rubbing material, and chemical, laser or electrocautery pleurodesis[8].

Global recurrence rates ranges from zero to 10%[8]. Many of these recurrences are due to failures of the method of treatment. Clipping, ligation and loop of blebs were associated with a recurrence of 11 - 22%[8]. The procedure of choice is stapling of blebs and bullae, or wedge resection of the tip of the lung when lesions are not identified, and pleural symphysis procedure such as mechanical abrasion, apical pleurectomy or patchy electrocoagulation of the parietal pleura. These procedures result in a recurrence rate of 0 - 5%[3-5,8]. In an uncontrolled series of 94 patients, the recurrence rate was 3.1% with a mean follow-up of 48 months[6].

Pleural procedures alone are associated with higher recurrence rates[8]. One reason for recurrence is failure to recognize the site of the air leak in the absence of blebs. Unrecognized blebs or inadequate resection of the diseased portion of the lung may also contribute. Another factor is inadequate pleurodesis, especially in between the tracer sites. These failures suggest that gauze pleural abrasion is probably less effective than apical pleurectomy.

Most recurrences occurred within the first month of the operation. Long-term follow-up did not add to the rate of recurrence[4].

Treatment of recurrences varies according to the size of pneumothorax and the presence of air leaks after insertion of chest tubes. For limited pneumothorax in stable patients, rest and observation are recommended. When reoperation is necessary, both repeat VATS and thoracotomy have been performed. In 50% of cases, residual bullae or air leaks are found; these are usually stapled and pleural symphysis procedure is added. When no blebs are identified, symphysis pleurodesis is performed[8].

In conclusion, in all cases of PSP, pathomorphologic changes were observed.

Even when no apical blebs or bullae are identified, pathology of the resected apex virtually always identifies paraseptal emphysema on such specimens. The actual site of air leakage was seen in some patients at VATS and at microscopic examination. VATS stapling of identified blebs or apex of the upper lobe and apical pleurectomy represent the standard treatment for recurrent or persistent PSP. In the absence of blebs or bullae, additional chemical pleurodesis might be indicated.

REFERENCES