Dear Editor,

We read with interest the article by Papagiannis et al., providing an introduction and categorization of the aetiology of pneumothorax into broadly primary and secondary (1). The aetiology of recurrent pneumothorax may deserve a special mention, although in the young population it is most likely due to primary cause from apical blebs or bullae, one should have a low threshold to perform further investigations to seek potential alternative pathological explanations for the recurrent spontaneous pneumothorax. We would like to share our experience of some rarer causes for recurrent pneumothorax.

Our thoracic surgical unit was previously referred two interesting cases of young men who presented with recurrent “primary” spontaneous pneumothorax for video-assisted thoracic surgery (VATS) pleurodesis. During thoracoscopy, the first patient had multiple cystic lesions of variable size over his entire lung (Figure 1). VATS resection of the lesions showed histological features of cystic formation, vascular congestion, interstitial hemorrhage and siderophage aggregation. There were fascicular spindle cells surrounding the cysts which stained positively for HMB 45 (Anti-Human Melanosome stain), smooth muscle actin (SMA) and desmin, hence to our surprise, confirming the diagnosis of pulmonary lymphangioleiomyomatosis (LAM). Classically, LAM is a disease limited to females of childbearing age, however, very rare cases of LAM in males have been described. When combining the reported cases and series in the literature, the female to male ratio for LAM is thought to be around 30 to 1, and this was indeed the first encounter of LAM in a male patient at our institute.

The second young gentleman presented with his second episode of “primary” spontaneous pneumothorax within a two-month period. He had no other past medical history. VATS pleurodesis was planned due to persistent airleak from his drain, however, during history taking, he divulged that he has been suffering from right thigh pain for a few months. Examination showed a deep mid-thigh lesion, suspicious of soft tissue tumour. Computed tomography of thorax revealed multiple bilateral cystic lung lesions with thin smooth walls of varying sizes (Figure 2). VATS exploration showed the airleak to be from a ruptured “cyst”, and resection of the lesion showed histological features of metastatic pulmonary epithelioid sarcoma.

The cases illustrate that clinicians should have a high index of suspicion for more sinister secondary causes of spontaneous pneumothorax in young men. As Forrest Gump (fictional character, 1994 film Forrest Gump, Paramount Pictures) once said, “Life is like a box of chocolates, you never know what you’re going to get”.

Yours sincerely,
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References