In 2015, a 63-year-old non-smoking male with a history of bullous emphysema, for which a wedge resection of the right lower lobe had been performed in 1972, presented with progressive dyspnoea on exertion and coughing. Pulmonary function tests demonstrated a moderate obstructive pattern with normal diffusion capacity (FEV₁: 61%, FVC: 64%, TLco: 86%). CT of the thorax revealed localised bullous-cystic changes in the right lower lobe with displacement of the mediastinum to the contralateral side (figure 1). Due to the patient’s increasing respiratory distress, a right lower lobectomy was performed.

Grossly, the specimen displayed multiple cysts with compression of the adjacent parenchyma (figure 2A). Histologically, placenta-like papillary structures characterised by clear cell proliferation admixed with fat tissue were observed. The clear cells expressed vimentin and CD10 but were negative for HMB-45. This histological pattern was consistent with placental lung transmogrification (PLT) (figure 2B, C). Based on these findings, the histology of the resected specimen from 1972 was revised. This specimen revealed an interstitial proliferation of CD10-positive clear cells, whereas placental-like papillary formations and generalised emphysematous changes were lacking (figure 2D), suggesting that the currently diagnosed PLT evolved from a progressively growing bullous-like lesion from 1972.

PLT is regarded as a rare variant of bullous emphysema that mainly affects young to middle-aged subjects. Clinical symptoms include dyspnoea, coughing, pneumonia, chest pain and...
Due to the slowly progressive nature of the disease, PL T may lead to functional impairment and adverse events, including infections or, as in the present case, compression of the contralateral lung due to mediastinal displacement. Surgical treatment of PL T in the form of lobectomy is adequate and effectively eliminates the risk of recurrent infections and improves lung function.

From a clinicopathological point of view, the differential diagnosis of PL T includes congenital pulmonary airway malformation (CPAM), bullous emphysema, fibrochondromatous hamartoma and lymphangioleiomyomatosis. However, lymphangioleiomyomatosis is improbable in this case as this disease is almost exclusively found in female patients. In the present case, histology, especially the lack of an interstitial clear cell component, was inconsistent with CPAM and emphysema. The clear cell proliferation mimics perivascular epithelioid cell-related disorders, including lymphangioleiomyomatosis or clear cell 'sugar' tumours, whereas from the clinical–radiological presentation, the characteristic histology and CD10 expression in the absence of HMB-45 immunoreactivity favour PL T. Histologically, hamartoma may simulate PL T because it sometimes contains fat tissue and displays a pseudopapillary, placental villi-like growth pattern. However, hamartomas are well-circumscribed masses that lack interstitial clear cells. It remains to be established whether the lipomatous change observed in PL T is attributable to metaplastic mesenchymal differentiation of the clear cell component.

To the best of our knowledge, this is the first case of progressing PL T with clear cell proliferations found in two consecutive resections. Cavazza et al. identified genotypical alterations in the clear cell component only, but not in the adjacent normal tissue. These data suggest that clear cells play a pivotal role in the pathogenesis and progression of PL T and support the concept that this rare disease cannot be interpreted as a variant of emphysema despite the striking clinical–radiological similarities.

REFERENCES