Diffuse idiopathic pulmonary neuroendocrine cell hyperplasia: a rare cause of airflow limitation

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DESCRIPTION

A woman in her 60s with end-stage renal disease presented to our respiratory medicine service following an incidental finding of pulmonary nodules and diffuse pulmonary mosaic attenuation on chest CT (figure 1), performed for workup of renal transplantation.

Apart from a mild chronic cough, the patient had no significant symptoms and had never smoked. Spirometry revealed moderate airflow limitation (FEV1 0.98 L: 50% predicted; FVC 1.46 L: 63% predicted) with no reversibility. Although the patient refused lung biopsy, the chest CT findings in addition to spirometry suggested the diagnosis of diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH).

DIPNECH is a rare disease attributed to the hyperproliferation of airway epithelial pulmonary neuroendocrine cells and peribronchiolar fibrosis resulting in airway narrowing. The cellular proliferation may lead to nodular densities called carcinoid tumourlets (≤5 mm) or tumours (>5 mm). DIPNECH is commonly seen in middle-aged non-smoking women. It can be discovered incidentally on imaging. Symptomatic patients manifest with dyspnoea, cough, wheezes and spirometry revealing airflow limitation. Radiological imaging is of paramount importance in establishing the diagnosis, and a CT scan typically reports mosaic attenuation related to air trapping and multiple solid pulmonary nodules. Other causes of irreversible airway obstruction are not associated with widespread pulmonary nodularities as seen in this case. The gold standard modality to diagnose DIPNECH is surgical lung biopsy with the histopathological demonstration of pulmonary neuroendocrine cells proliferation.

The prognosis is variable; patients can remain stable while some may progress slowly to respiratory failure due to constrictive bronchiolitis. Because of the rarity of the disease, there is no established treatment. Close observation is usually preferred for asymptomatic patients. Pharmacological therapy for symptomatic patients includes inhaled bronchodilators, steroids, somatostatin analogues and sirolimus, with varying success.

Learning points

► Although rare, DIPNECH should be considered in the differential diagnosis of irreversible airflow limitation.
► In DIPNECH patients, CT chest typically reveals pulmonary nodules and evidence of air trapping with mosaic attenuation.

Figure 1 (A) Axial CT image of the chest below the level of the carina shows mosaic lung attenuation, with geographical areas of decreased attenuation (asterisk) adjacent to normal lung and multiple bilateral scattered small (<5 mm) solid lung nodules, corresponding to tumourlets (arrows). (B,C) Maximum intensity projection images at the same level clearly demonstrate the bilateral small lung nodules (arrows) within the areas of air trapping. (D) Minimum intensity projection image demonstrates better visualisation of the mosaic lung attenuation and air trapping.

REFERENCES
