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PULMONARY PUZZLE

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Answer

Transthoracic needle aspiration of the mass showed abundant small mature lymphocytes and some atypical cells with cytological abnormalities. An immunoperoxidase panel revealed a positive reaction with pankeratin, vimentin and LCA stains. Stains for MOC-31, Ber-EP4, TTF-1 and calretinin were negative. The pathologist at the local hospital suggested a possible large cell carcinoma of a lymphoepithelioma type but requested further investigation.

A CT-guided core biopsy stained for cytokeratin 5/6 and p63, supporting the diagnosis of a pleural-based thymoma. There was a separate infiltrate consistent with lymphocytic interstitial pneumonitis (LIP). This was confirmed by a video-assisted thoracoscopic surgical lung biopsy (fig 1). The diagnosis of a pleural-based thymoma was made in the absence of a mediastinal mass on initial imaging.

The thymoma was treated with six cycles of cisplatin and etoposide, with prednisone for the LIP. Follow-up imaging 8 months later showed significant improvement in both the pleural thymoma and LIP.

Pleural thymoma

Thymomas are epithelial-type neoplasms that typically occur in the anterosuperior mediastinum. Up to 4% of thymomas originate from ectopic thymic tissue.[1 2] Diagnosis of a pleural-based thymoma is difficult, with the differential diagnosis including lymphoma, mesothelioma, sarcoma and other metastases or spread from a primary pulmonary malignancy.

The course of pleural thymoma is usually indolent with non-specific symptoms. Imaging can demonstrate a unifocal mass, multifocal masses, diffuse pleural thickening or pleural effusions. Primary pleural thymoma does not have the mediastinal involvement usually seen in lymphoma. Histological examination is required for the diagnosis and has recently been reviewed.[2] Briefly, thymomas consist of thymic epithelial cells incompletely surrounded by fibrous septae with infiltrating polyclonal lymphocytes. Immunohistochemistry can distinguish thymomas from other malignancies. Cytokeratin stains cells derived from an epithelial lineage, including thymoma and mesothelioma. Thymomas typically stain positive for cytokeratin 5/6, p63, CD99, CD1a and TdT.[2]

Our patient had a bilateral pulmonary infiltrate consistent with LIP. While thymomas often have a polyclonal lymphocytic infiltrate, they do not typically cause a diffuse pulmonary infiltrate as seen in this case. To our knowledge this is the first case of a thymoma-associated LIP.

From the question on page 666

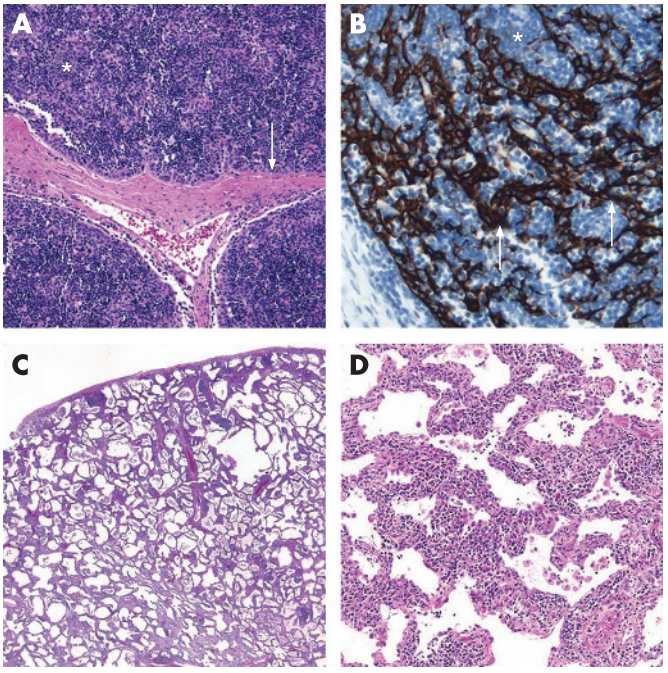

Figure 1 (A) Photomicrograph of thymoma demonstrating the typical features of a lymphoid and epithelial proliferation (*) traversed by hyalinised fibrous septae (arrow). H&E; 40× original magnification. (B) Medium power photomicrograph of the thymoma in which pankeratin immunohistochemistry (dark stain, arrows) highlights the network of neoplastic thymic epithelial cells interspersed within a benign lymphoid stroma (*). 100× original magnification. (C) Low power scanning photomicrograph of the lung wedge biopsy demonstrating diffuse interstitial infiltration of lymphocytes and plasma cells. H&E; 12.5× original magnification. (D) Medium power photomicrograph of lung wedge demonstrating expansion of the alveolar septae by mononuclear inflammatory cells. 110× original magnification.

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