
PULMONARY PUZZLE

Answer
Thoracotomy demonstrated non-casing epithelioid cell granulomas with focal central coagulative necrosis in pulmonary and pleural specimens (fig 1C). Periodic acid Schiff, alcin blue, methenamine silver stains for fungi, Ziehl-Neelsen stain, culture for Mycobacterium tuberculosis and non-tuberculous mycobacteria were negative. The final diagnosis was pleuropulmonary sarcoidosis.

The non-compromised youth was clinically characterised with a protracted tolerable fever of unknown origin and diffuse pleural thickening with multifocal pulmonary micronodules. The clinical presentation and laboratory examinations did not suggest non-specific bacterial infections or collagen vascular diseases as causes of the fever.

Although the patient displayed some radiological features of malignant pleural mesothelioma (fig 1A), this usually develops in older patients and presents with pleuritic chest pain, large pleural effusion and rind-like pleural thickening.

A negative tuberculin skin test and diffuse pleural thickening without a pleural effusion did not suggest tuberculous pleurisy.

Thoracotomy with histological examination identified pleuropulmonary sarcoidosis as the cause of the fever of unknown origin. With focal central necrosis, strenuous efforts were taken to rule out fungal and mycobacterial infections.

As a multisystem inflammatory disorder, sarcoidosis could present as dyspnoea and fever with multiorgan involvement. In Scadding β sarcoidosis, intrathoracic adenopathy is absent while perivascular or subpleural micronodules and bronchovascular bundle thickening are characteristic features on the HRCT scan, corresponding to the spirometric finding of impaired pulmonary transfer factor.

The absence of bilateral hilar adenopathy and hypercalcemia and a normal ACE level might be associated with the delay in diagnosis. In fact, ACE is no longer listed in the diagnostic criteria for sarcoidosis and only 5% of patients with sarcoidosis have hypercalcemia.

Clinically significant pleural thickening in sarcoidosis due to pleural infiltration of sarcoid granuloma is rare and usually occurs in patients with advanced stage disease who present with chest pain and/or dyspnoea and scarce pleural effusion. In our case, prednisone 40 mg/day resolved the fever and alleviated the dyspnoea 7 days after starting treatment.

Sarcoidosis could present as diffuse pleural thickening with mediastinal pleural involvement, especially in the advanced stage. Some patients might show a good response to corticosteroids. Sarcoidosis should be listed in the differential diagnosis of patients with fever of unknown origin. A normal ACE level, the absence of hilar adenopathy and hypercalcemia might prolong the diagnosis.

From the question on page 622.

REFERENCES