Clinics in Diagnostic Imaging (27)

A N Leung

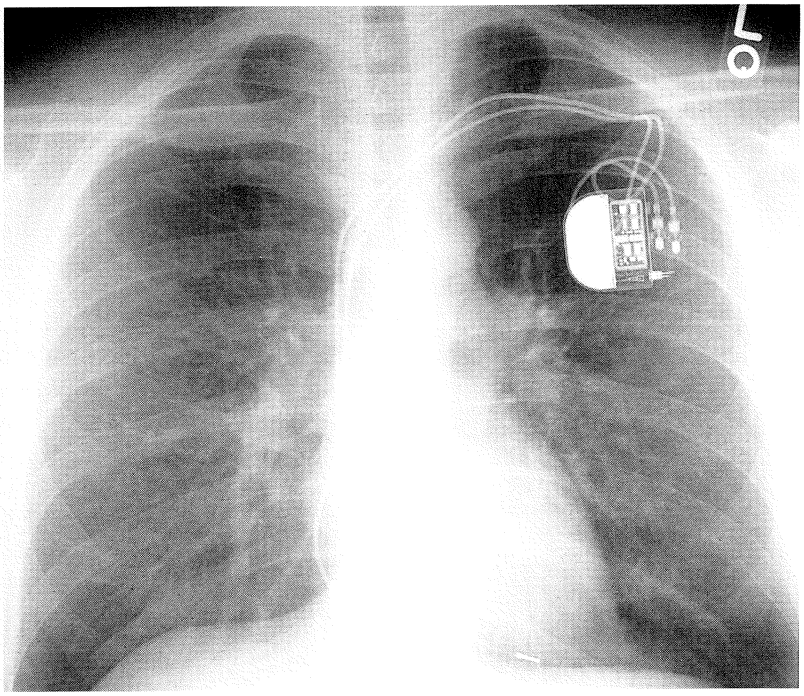

Fig 1 - PA chest radiograph

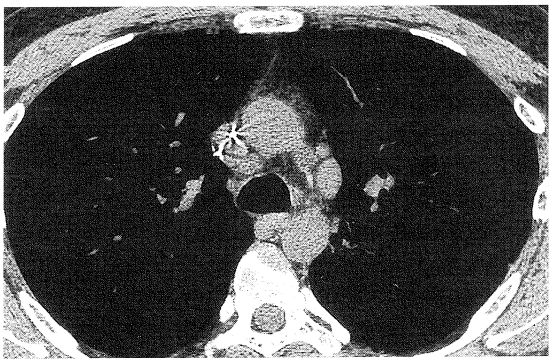

Fig 2A - 1mm HRCT scan at level of aortopulmonary window displayed at mediastinal settings.

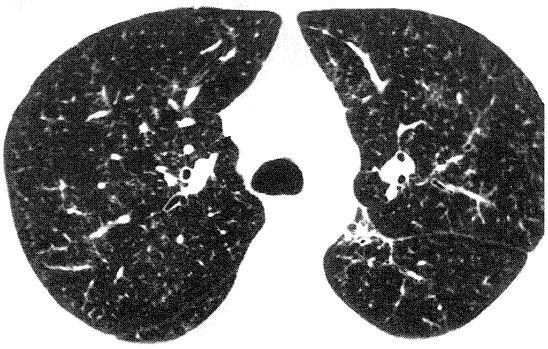

Fig 2B - Corresponding 1mm HRCT scan displayed at parenchymal settings.

CASE REPORT

A 44-year-old Caucasian man presented with symptoms of weakness and decreasing exercise tolerance. An electrocardiogram demonstrated a third-degree heart block which was treated with placement of a dual chamber pacemaker.

Posteroanterior chest radiograph (Fig 1) and computed tomography (CT) of the thorax (Fig 2) were performed. What do these demonstrate? What further investigations should be performed for confirmation of the diagnosis?

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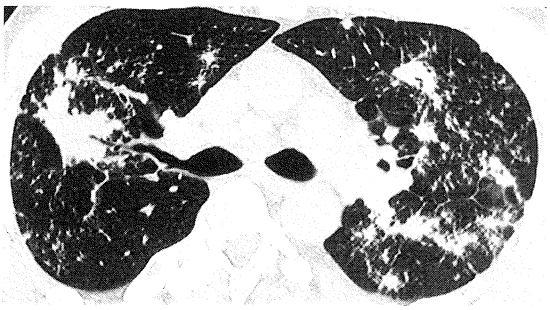

Fig 3 - 1.5 mm HRCT scan of the upper lobes shows multiple, bilateral nodules of varying sizes. Note the presence of an air bronchogram in the right upper lobe conglomerate mass.

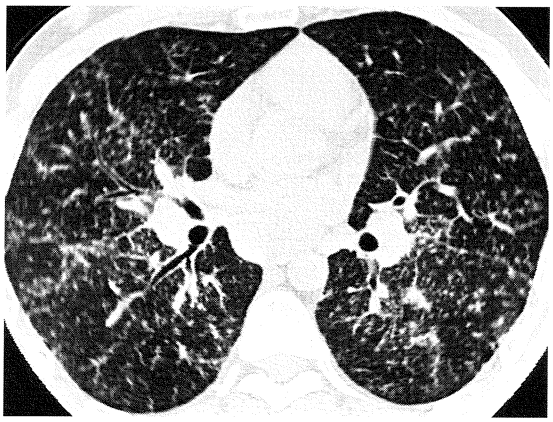

Fig 4 - 1.5 mm HRCT scan of 26-year-old Caucasian man with sarcoidosis shows bilateral hilar lymphadenopathy and micronodules distributed along perilymphatic routes, causing nodular thickening of peribronchovascular interstitium, interlobular septa, and subpleural regions.

IMAGE INTERPRETATION

Chest radiograph (Fig 1) showed right paratracheal, aortopulmonary window, and subtle bilateral hilar lymphadenopathy associated with a diffuse reticulonodular parenchymal pattern. CT confirmed the presence of mediastinal (Fig 2A) and bilateral hilar lymphadenopathy (not shown); the diffuse micronodular pattern predominated in the middle and upper lung zones (Fig 2B).

DIAGNOSIS

Sarcoidosis

CLINICAL COURSE

After placement of the pacemaker, all of the patient's symptoms resolved. Specifically, he denied fevers or any residual cardiopulmonary symptoms. His measured angiotensin converting enzyme (ACE) level was normal. Transbronchial biopsies were non-diagnostic. In order to justify treatment with corticosteroids, definitive diagnosis was obtained via mediastinoscopy. Histopathologic examination of biopsied lymph nodes revealed the presence of non-caseating granulomas consistent with sarcoidosis.

DISCUSSION

Sarcoidosis is a multisystemic granulomatous disorder of unknown aetiology that involves the lungs in 90% of patients[1]. Although thoracic manifestations are most common, virtually any organ system may be affected[2]. Ocular, neurologic, and cardiac involvement usually require prolonged and high dose of corticosteroid treatment[3].

Thoracic sarcoidosis may present with a myriad of radiographic findings ranging from normal (Stage 0) to parenchymal fibrosis (Stage IV)[4]. The most characteristic radiographic finding is lymphadenopathy which is present in approximately 50% of patients[4]. Radiologic accuracy and confidence in the diagnosis of thoracic sarcoidosis has been improved by evaluation using high-resolution computed tomography (HRCT)[5].

On HRCT, nodules are the predominant parenchymal lesion in sarcoidosis[6]. Nodules may range in size from a few mm to several cm; are typically irregular in margination, and have a very distinctive distribution – along bronchovascular bundles, interlobular septa, and subpleural regions – reflecting the pathologic distribution of sarcoidosis granulomas along lymphatic pathways[7]. Sarcoidosis nodules are found predominantly in the upper and middle lung zones. Intralobular reticular opacities, architectural distortion, and traction bronchiolectasis/ bronchiectasis are seen in patients who have progressed to fibrosis.

Occasionally, the predominant parenchymal finding in sarcoidosis may consist of single or multiple large nodules (> 1 cm), masses, or mass-like areas of consolidation with cavitation or air-bronchograms (Fig 3). Histologically, these findings represent aggregations of sarcoidosis granulomas sometimes present in association with surrounding fibrosis[8].

In the differential diagnosis, sarcoidosis (Fig 4) must be distinguished from two other nodular diseases – lymphangitic carcinomatosis (LC) (Fig 5) and lymphoma (Fig 6) – that also have a perilymphatic distribution and are often associated with intrathoracic lymphadenopathy. Often the clinical context, particularly if a previous history of malignancy can be elicited, can help prioritise the differential possibilities. Sarcoidosis is one of few pulmonary

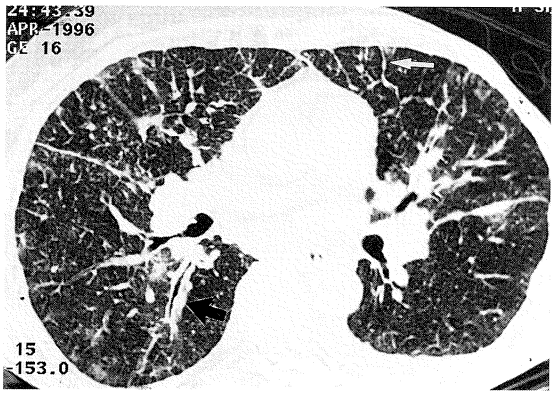

Fig 5 - 1.5 mm HRCT scan of 71-year-old woman with previous history of breast carcinoma and newly-diagnosed lymphangitic carcinomatosis shows bilateral hilar lymphadenopathy and nodular thickening of right lower lobe peribronchovascular interstitium (large arrow). In comparison to the case of sarcoidosis illustrated in Fig 4, there is a more prominent reticular (linear) pattern with better delineation of interlobular septal (small arrow) and fissural thickening.

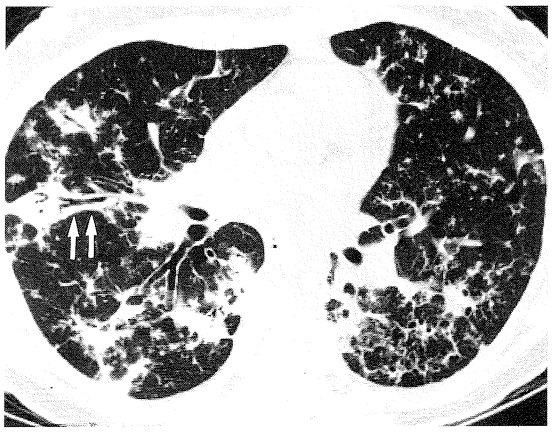

Fig 6 - 1.0 mm HRCT of 48-year-old woman with Hodgkin's lymphoma shows bilateral hilar lymphadenopathy. Poorly-marginated nodules are scattered bilaterally associated with peribronchovascular consolidation (arrows).

disorders in which the radiologic extent of disease may be much more impressive than the patient's symptomatology. On HRCT, although all three of the previously mentioned diseases may cause interlobular septal thickening, formation of well-defined "polygonal arcades" occurs more frequently in the neoplastic diseases as does focal disease restricted to one lobe or one side. Peribronchovascular consolidation (Fig 6) is a characteristic feature of lymphoma and may be so extensive as to mimic a pneumonia. HRCT findings of fibrosis (architectural distortion, traction bronchiectasis, intralobular reticular opacities, honeycombing) would favour sarcoidosis as it does not occur in either LC or lymphoma.

In some patients, as exemplified by the reported case, the perilymphatic distribution of sarcoid nodules may not be apparent on HRCT. The differential diagnosis of disseminated micronodules (< 7 mm in size) with associated lymphadenopathy includes pneumoconioses, miliary dissemination of infectious granulomatous diseases including tuberculosis, lymphoma, and metastatic disease from thyroid carcinoma, renal cell carcinoma, or poorly differentiated adenocarcinoma from any site. Again the clinical context, particularly with respect to exposure history or acuteness of illness, may be critical in appropriately narrowing the differential possibilities.

The diagnostic criteria for sarcoidosis are well established and consist of: 1) compatible clinical or radiologic evidence or both; 2) histologic evidence of noncaseating granulomas, and 3) negative bacterial and fungal studies of biopsied tissue or sputum[9]. Although radiologic findings may be consistent or highly suggestive of the diagnosis, they are not pathognomonic and it is generally felt that if treatment with corticosteroids is indicated, the diagnosis should be confirmed with histology[10]. Additionally, tissue cultures should always be performed to exclude an infectious granulomatous disease.

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ABSTRACT

A 44-year-old Caucasian man presented with third-degree heart block. Chest radiograph and high-resolution computed tomography (HRCT) of the thorax showed mediastinal and bilateral hilar lymphadenopathy associated with a diffuse, bilateral micronodular pattern. The HRCT findings and differential diagnosis of sarcoidosis are reviewed.

Keywords: computed tomography, high-resolution; sarcoidosis