Review Article

Pulmonary sarcoidosis: high-resolution computed tomography findings*

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Sarcoidosis is a systemic disease of unknown etiology, characterized by noncaseating granulomas. Although it may affect any organ, morbidity and mortality are most commonly related to pulmonary involvement, which is found in 80-90% of patients. This study illustrates the principal manifestations of sarcoidosis seen in high-resolution computed tomography scans, including typical as well as atypical forms.

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Key words: Sarcoidosis. Lung. High-resolution computed tomography.

INTRODUCTION

Sarcoidosis is a multisystemic disease of unknown etiology that is variable in its presentation, progression and prognosis. Pulmonary involvement is seen in up to 90% of patients, and 20-25% of those present permanent functional impairment⁽¹⁻⁴⁾.

Pulmonary involvement tends to be bilateral and asymmetric, predominantly found in the upper lobes. Radiologic findings are atypical in 25% of the cases, and chest X-rays reveal normal results in 5-10% of patients^(5,6).

The objective of this study was to present, in a succinct and illustrative way, the main aspects of

pulmonary sarcoidosis revealed in high-resolution computed tomography scans.

TYPICAL FINDINGS

Nodules. The nodular pattern is the pattern most frequently seen in pulmonary sarcoidosis. The nodules are generally small and present perilymphatic distribution, involving the peribronchovascular cuffs, interlobular septa, subpleural region, centrilobular areas and the entire length of the fissures (Figures 1 to 3). Nodules may be large or cavitary, sometimes mimicking neoplasms, and are found in 15-25% of the cases (Figure 4)^(1,4,7-10).

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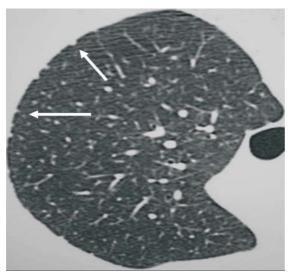


Figure 1. Micronodular form. Micronodules in the subpleural regions (arrows)

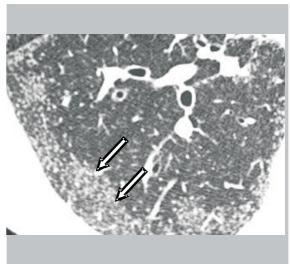


Figure 3. Micronodular form. Confluent peripheral micronodules (arrows)

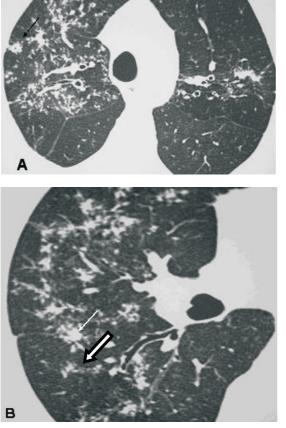


Figure 2 A and B. Micronodular pattern. Micronodules in the upper lobes, some of which are confluent (arrows)

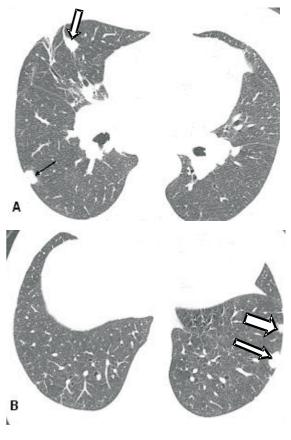


Figure 4. Nodular form. Axial images of the middle third (A) and base (B) of the lungs, revealing multiple peripheral nodules of various dimensions (arrows)

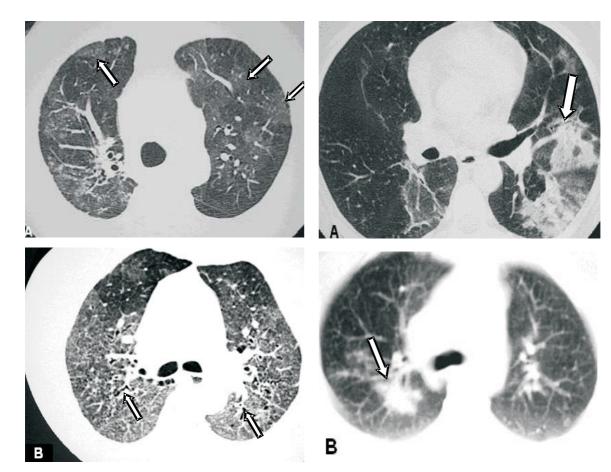


Figure 5. Ground-glass pattern. A – Scattered bilateral ground-glass opacities (arrows). B –Ground-glass pattern accompanied by traction bronchiectasis (arrow)

Figure 6. Parenchymal opacities A – Poorly defined opacities in the left lung, accompanied by air bronchograms (arrow). B – lrregular consolidation in the right lung (arrow)

Ground-glass opacities. Ground-glass opacities are found in 14-83% of patients, tending to present scattered distribution. It may occur in isolation or be accompanied by signs of pulmonary fibrosis (Figure 5)⁽¹¹⁻¹³⁾.

Parenchymal opacities. Parenchymal opacities are less common than ground-glass opacities and are generally seen in the initial stage of the disease. They can be found throughout the peribronchovascular cuffs or on the lung periphery. Their borders are irregular, and this type of opacity is frequently accompanied by air bronchograms and nodules in the adjacent parenchyma (Figures 6 and 7)⁽¹⁴⁾.

Reticular opacities. Reticular opacities result from the combination of the thickening of interlobular septa and of the intralobular interstice. Septal

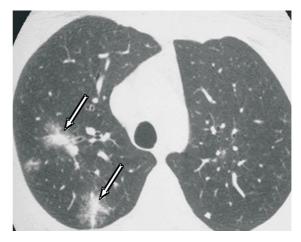


Figure 7. Parenchymal opacities with imprecise, sparse borders in the right upper lobe, surrounded by ground-glass pattern (arrows)

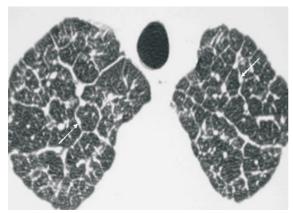


Figure 8. Reticular opacities. Pulmonary sarcoidosis with septal thickening of the upper lobes, forming polygonal arches (arrows), simulating lymphangitic carcinomatosis

thickening is typically found along the peribronchovascular cuffs but can also be seen in other topographies (Figure 8). Irregular linear opacities adjacent to the bronchovascular cuffs have also been described and attributed to early manifestations of pulmonary fibrosis (Figure 9)^(7,9).

Air trapping. Air trapping during expiration is relatively frequent, due to the presence of peribronchial or submucosal granulomas or to peribronchiolar fibrosis, leading to small airway obliteration (Figure 10) (15).

Fibrosis. Parenchymal abnormalities may evolve to fibrosis, which is accompanied by architectural distortion, volumetric loss, linear opacities, honeycombing, fibrotic masses, bronchiectasis and traction bronchiolectasis. Fibrosis is usually found in the upper and middle lung fields (Figures 11 and 12)⁽⁷⁾.

Bronchiectasis. Bronchiectasis is an uncommon finding in sarcoidosis and is frequently associated with fibrosis (traction bronchiectasis). Other less common etiologies are airway obstruction caused by granulomas and extrinsic compression caused by lymph nodes (Figure 13)^[6].

ATYPICAL FINDINGS

Pseudotumoral form. In some cases, coalescence of interstitial granulomas results in large conglomerates with the formation of masses (pseudotumors not accompanied by air bronchograms) that compress adjacent air spaces (Figures 14 and 15)⁽¹⁶⁾.



Figure 9. Reticular opacities. A – Irregular thickening of peribronchovascular cuffs (arrows). B – thickening of posterior basal interlobular septa (arrow) accompanied by distortion of the lung architecture

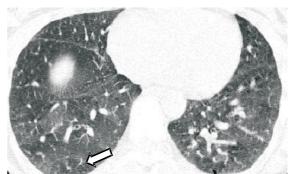
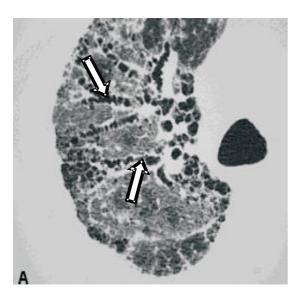
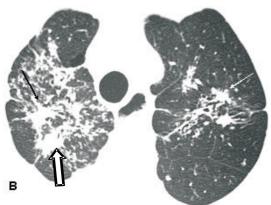


Figure 10. Areas of air trapping in the lower lobes (arrows)

Cysts. The etiology of cysts is uncertain. Of all the possible causes, peripheral air imprisonment, alveolar distention caused by endobronchial component, the destruction of the alveolar parenchyma, and retractions and collapses of the surrounding parenchyma have been reported (Figure 16)⁽⁷⁾.

True cavities. True cavities are extremely rare. Infectious causes and massive fibrosis with cysts must





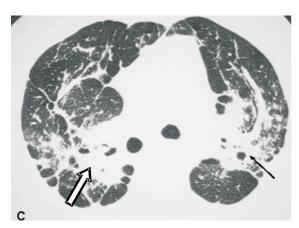


Figure 11. Fibrosis. A – Traction bronchiectasis (arrows), architectural distortion and ground-glass pattern, showing the classical aspect of fibrosis. B – Irregular parenchymal opacities accompanied by architectural distortion (arrows). C – Fibrotic conglomerate forming masses with retractile aspect (arrows)

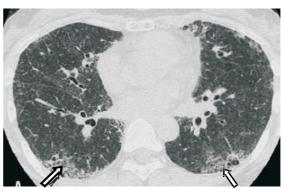


Figure 12. Fibrosis. Posterior subpleural honeycombing, predominantly in the lower lobes (arrows)

first be ruled out. True cavities, as well as apical cysts, may be the focus of saprophytic colonizations^(17,18).

Bronchial changes (thickening/stenosis). Lenique et al. reported thickening of the bronchial walls, caused by the deposition of granulomas along the peribronchovascular interstice, in 65% of patients (Figure 17)⁽¹⁹⁾.

Lobar atelectasis. The occlusion of a lobar bronchus and consequent atelectasis may be caused by intraluminal disease or extrinsic compression. There are no radiological signs that help distinguish between lobar atelectasis caused by sarcoidosis and that resulting from other causes⁽¹⁷⁾.

Unilateral pulmonary parenchymal lesions. It has been reported in the literature that unilateral parenchymal changes are caused mainly by localized interstitial disease, accompanied by an alveolar pattern, or by localized interstitial disease, accompanied by a reticulonodular pattern or presenting as a pulmonary coin lesion^(7,17).

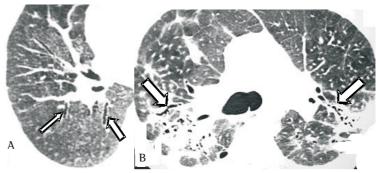


Figure 13A. Bronchiectasis (arrows) associated with peripheral micronodules. B – Irregular fibrotic mass with traction bronchiectasis (arrows)

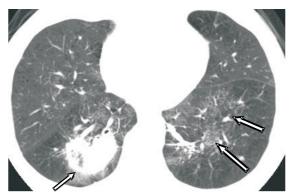


Figure 14. Pseudotumoral form. Mass with irregular borders in the right upper lobe (arrow). Ground-glass areas characterized in the left lower lobe and adjacent to the mass (arrows)

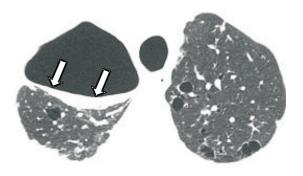


Figura 16. Sarcoidose com cistos bilaterais associados a fístula broncopleural com formação de hidropneumotórax à direita (setas)

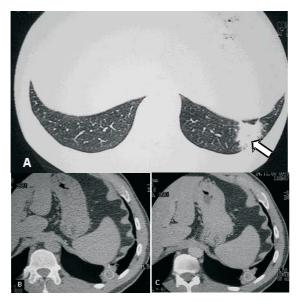


Figure 15. Pseudotumoral form. A – High-resolution computed tomography with lung window showing spiculated mass in the posterior basal segment of the left lower lobe (arrow). B – Density measurement during the pre-contrast phase (40 UH). C – Scan three minutes after intravenous administration of iodinated contrast material, highlighting the lesion (maximum density of 76 UH). Swensen's protocol positive(16). Anatomopathological examination of the surgical sample confirmed sarcoidosis

Fairy-ring sign. The "fairy-ring sign" is caused by multiple peripheral granulomatous nodules bordering areas of preserved lung parenchyma (Figure 18)⁽²⁰⁾..



Figure 17. Bronchial involvement. Unilateral presentation, characterized by concentric thickening of the bronchial walls (arrows), with nodules with lymphatic distribution in the right lower lobe

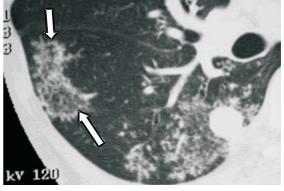


Figure 18. Fairy-ring sign. Micronodules with ring-like distribution (arrows) surrounding relatively preserved lung parenchyma

CONCLUSIONS

Because of the broad spectrum of presentations, pulmonary sarcoidosis may mimic other interstitial pulmonary diseases. High-resolution computed tomography is the most sensitive and specific imaging method for the evaluation of this disease.

In those cases in which atypical manifestations predominate, the knowledge of the various tomographic patterns helps us limit the differential diagnosis and select the best location for an eventual biopsy.

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