

INTRATHORACIC SARCOIDOSIS: ATYPICAL RADIOLOGICAL FORMS FOUND IN CLINICAL PRACTICE

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ABSTRACT

Sarcoidosis is a multisystemic chronic inflammatory disease characterized by noncaseous epithelioid cell granulomas. Involvement of the lung and of bilateral hilar and mediastinal lymph nodes is most common. Atypical patterns include unilateral lymphadenopathy, that can be seen in unusual locations or may become calcified.

The most frequent parenchymal pattern of sarcoidosis is a bilateral perilymphatic symmetric distribution of micronodular lesions. Miliary opacities or ill-defined irregular opacities, without air bronchograms, which develop cavitation, with satellite nodules are seen in other granulomatous diseases and neoplasms. Occasionally, parenchymal lesions coalesce, forming multiple well-defined macronodules mimicking a metastatic process.

Stenosis or compression of bronchi may result from endobronchial granulomas, resulting in obstruction and atelectasis. Pleural involvement is rare including pleural effusion, pneumothorax, pleural thickening or calcifications.

Thoracic sarcoidosis manifests with various patterns. Atypical manifestations of sarcoidosis may require an extensive differential diagnosis that encloses tuberculosis, silicosis, pneumoconiosis, malignancies,

RÉSUMÉ

La sarcoïdose intra-thoracique: formes radiologiques atypiques trouvées dans la pratique clinique

La sarcoïdose est une maladie granulomateuse systémique à localisation médiastino-pulmonaire prédominante. Elle est asymptomatique ou prend souvent des signes généraux: fatigue, sueurs nocturnes, toux, dyspnée. Elle est dite atypique lorsque certaines présentations clinique, radiologique ou évolutive de l'atteinte médiastino-pulmonaire ou des localisations extrathoraciques sont atypiques.

L'aspect radiologique atypique montre des images de l'atteinte miliaire parenchymateuse, des opacités alvéolaires bilatérales excavées (nécrobiose au sein de zones confluentes granulomateuses), des adénopathies médiastinales calcifiées, atteinte unilatérale, ganglionnaire ou parenchymateuse, atteinte pleurale (épaississements pleuraux ou pleurésie) ou formes ganglionnaire médiastinale et parenchymateuse d'un aspect pseudotumoral. La sténose d'allure tumorale est la forme atypique observée après l'endoscopie bronchique.

On rappelle quelques aspects inhabituels de la sarcoïdose (miliaire parenchymateuse, pleurésie, lâcher de ballons, aspergillome, fibrose pulmonaire, emphysème, suspicion d'une infection ou d'un cancer

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lymphoma, and any other causes of chronic pulmonary infiltrates.

Key words: sarcoidosis, atypical forms, mediastinal lymph nodes.

INTRODUCTION

Sarcoidosis represents a multisystem chronic inflammatory condition characterized by noncaseous epithelioid cell granulomas and changes in tissue architecture, that can affect any organ. Involvement of the lung and the hilar and mediastinal lymph nodes is most usual. (approximately 90% cases)¹

CLINICAL FEATURES

The most frequent clinical features are symptoms of the respiratory tract (dyspnea, cough, bronchial hyper-reactivity), night sweats, weight loss, fatigue and erythema nodosum. Nonetheless, roughly half of patients suffering of sarcoidosis are asymptomatic, abnormalities are determined remotely on the paraclinical investigations (chest radiography)².

Common early-stage features that are associated with a good prognosis are fever, polyarthritis, erythema nodosum, and bilateral hilar lymph node enlargement (Löfgren's syndrome)³. Factors associated with a poor prognosis include radiologic stage 2 or 3 pulmonary disease at the time of initial diagnosis, disease onset after the age of 40 years, black race, splenomegaly, hypercalcemia, chronic uveitis, osseous involvement, and lupus pernio⁴.

USE OF HIGH-RESOLUTION CT

High-resolution CT may be notably useful for differentiating active inflammation from irreversible fibrosis in chosen patients. Nodules, ground-glass or alveolar opacities are evocative of granulomatous inflammation that may be inverted with treatment⁵. In comparison, honeycomb-like cysts, bullae, architectural distortion, volume loss, and traction bronchiectasis are suggestive for irreversible fibrosis⁶. Nonetheless, atypical manifestations of sarcoidosis may require an extensive differential diagnosis that encloses tuberculosis and other granulomatous infections, pneumoconiosis, and neoplastic diseases⁷.

TYPICAL PATTERNS OF LYMPH NODE ENLARGEMENT

The most usually seen pattern is bilateral hilar and mediastinal lymph node enlargement (95%)³.

surajouté) et montrent les difficultés diagnostiques qu'elles posent.

Mots clés: sarcoïdose, formes atypique, adénopathies médiastinales.

Bilateral lymph node enlargement can predict a possible infection (fungal or mycobacterial) or malignancy (figure 1)⁸.

ATYPICAL PATTERNS OF LYMPH NODE ENLARGEMENT

In certain occasions, lymphadenopathies may be asymmetric (unilateral) or seen in locations that are unusual (retrocrural regions, internal mammary and paravertebral)⁷. Lymphadenopathies in some cases become calcified and this feature is most related to the course of the disease⁹. Atypical patterns of lymph nodes enlargement occur more common in elderly patients, over 50 years (figure 2)¹⁰.

TYPICAL PARENCHYMAL MANIFESTATIONS

Micronodular lesions with perilymphatic distribution is the most common parenchymal disease pattern seen in patients with sarcoidosis. These are symmetric and bilateral, prevalent in the upper and middle lobes, typically in the interstitium of both subpleural and peribronchovascular spaces. In around one-quarter of cases, fibrotic changes are developing specifically over time, producing CT findings like architectural distortion, traction bronchiectasis and linear opacities (figure 3)^{7,8,10}.

ATYPICAL PARENCHYMAL MANIFESTATIONS

Irregular ill-defined opacities measuring maximum 4 cm in diameter that may or may not express with air bronchograms or rarely develop cavitation, with small satellite nodules that are often seen at the periphery of these opacities, that accompany the lymph vessels, can be seen in the presence of neoplasms and other granulomatous diseases (figure 4)^{8,12,13}.

In some occasions, parenchymal opacities confluence, forming conglomerate lesions, that may appear like multiple well-defined rounded macronodules mimicking a metastatic process (figure 5)^{7,10}. The confluence of many micronodular opacities compressing the surrounding alveoli or encroaching the alveolar spaces may mimic cryptogenic organizing pneumonia, pulmonary tuberculosis or community pneumonia (figure 6)¹¹.

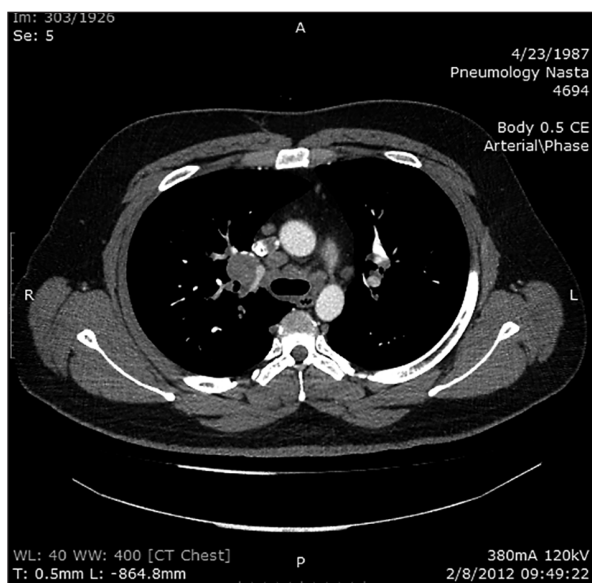


Figure 1. Typical radiological findings of lymphadenopathy in a patient with sarcoidosis. Axial contrast material-enhanced CT scan (mediastinal window) obtained at carinal level shows typical bilateral and symmetric hilar lymph nodes.

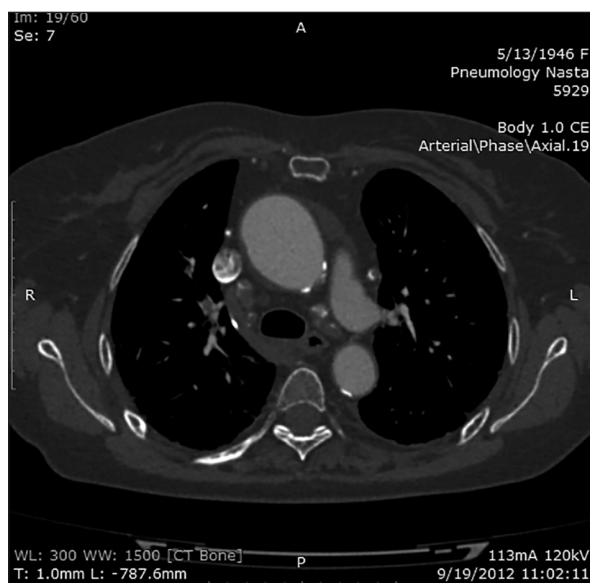


Figure 2. Atypical radiological findings of lymphadenopathy in a patient with sarcoidosis. Axial contrast-enhanced CT scan (mediastinal window) shows bilateral eggshell-like calcifications of hilar and mediastinal lymph nodes.

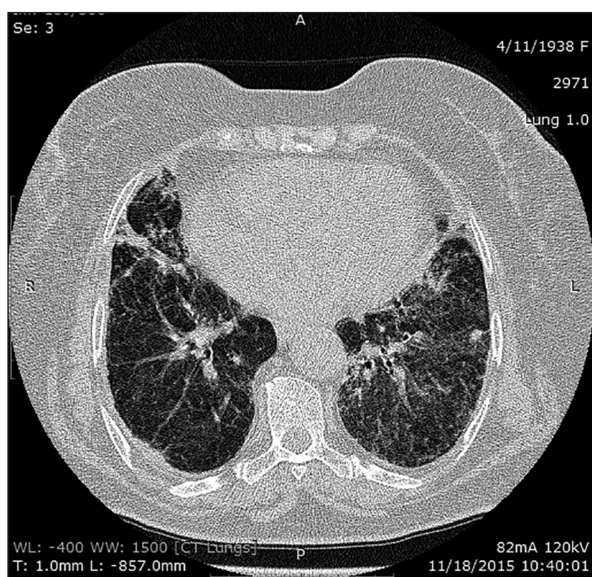


Figure 3. Axial high-resolution CT scan (pulmonary parenchymal window) in a woman with pulmonary sarcoidosis shows the typical perilymphatic distribution of micronodules.

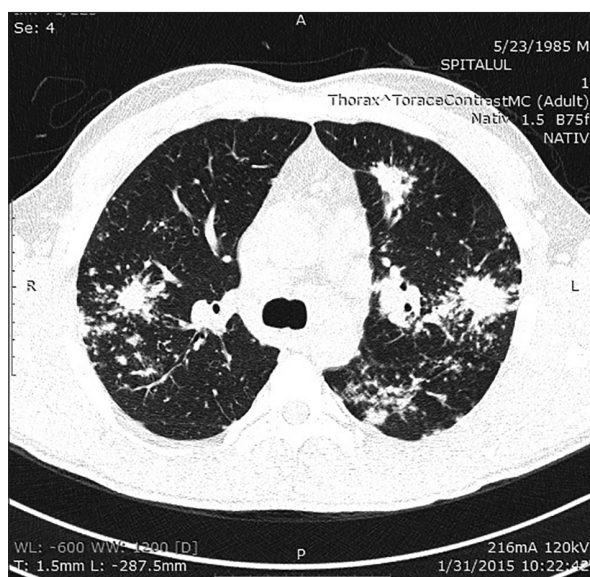


Figure 4. Axial high-resolution CT scan (pulmonary parenchymal window) shows several large, ill-defined nodules and areas of consolidation resulting from the confluence of multiple parenchymal micronodules in both lungs.

Patchy ground-glass opacities result from the coalescence of multiple micronodular granulomatous and fibrotic interstitial lesions (figure 7)³. Irregular intralobular and interlobular septal thickening it can be observed in the subpleural space and may simulate lymphangitic carcinomatosis¹¹.

Miliary opacities, a rare feature seen in sarcoidosis (less than 1% of patients), may guarantee

the inclusion of diseases such as pneumoconiosis, tuberculosis, histoplasmosis, chickenpox, metastatic lesions and Langerhans cell histiocytosis in the differential diagnosis (figure 8)¹⁴.

Honeycomb-like images seen in patients suffering from sarcoidosis are most often found in subpleural regions of the middle and upper lung zones, lung bases being usually spared (figure 9)¹⁵.

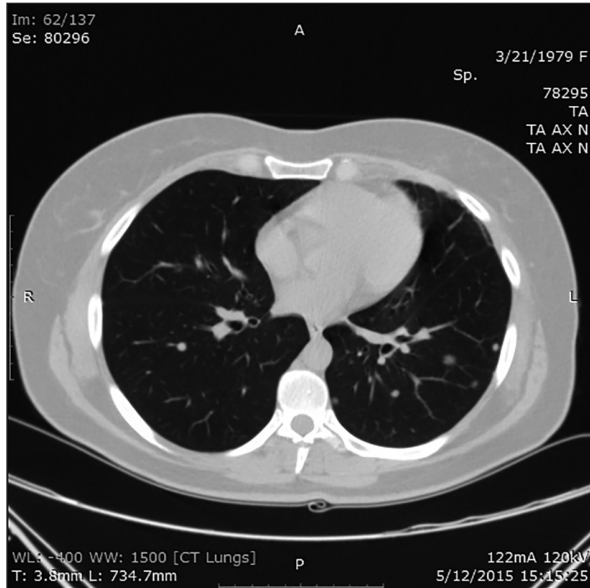


Figure 5. Contrast enhanced CT scans (pulmonary parenchymal window) show pulmonary nodules in subpleural and fissural regions. Pulmonary biopsy confirmed sarcoidosis.

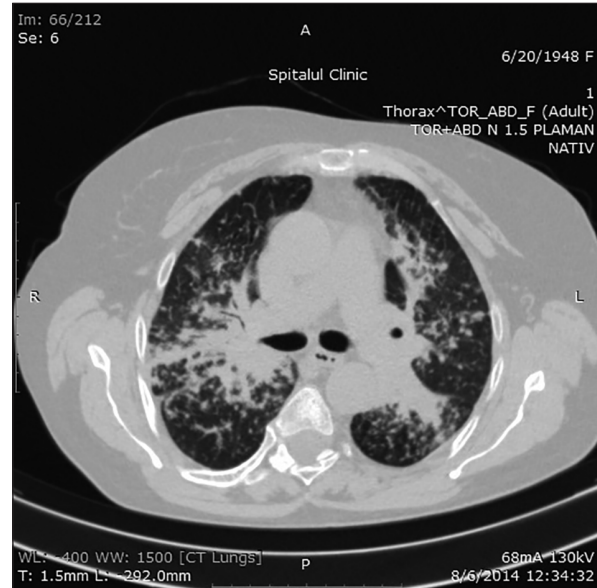


Figure 6. Axial high-resolution CT scan (pulmonary parenchymal window) shows atypical pattern in pulmonary sarcoidosis with alveolar consolidation, architectural distortion, traction bronchiectasis and signs of fibrosis.

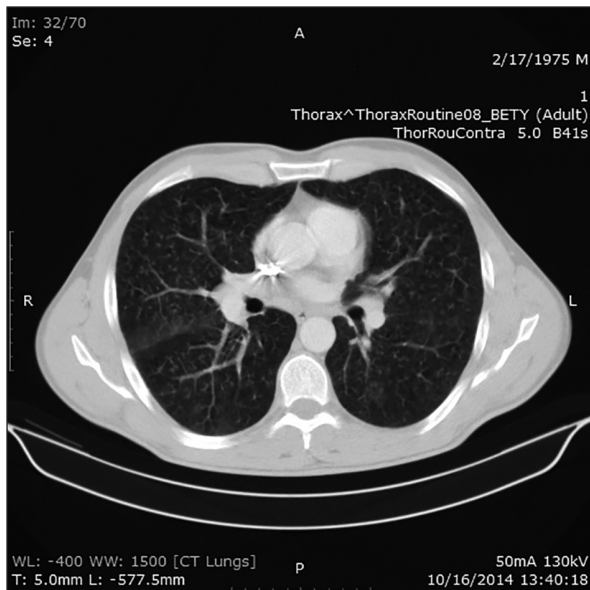


Figure 7. Axial high-resolution CT scan (pulmonary parenchymal window) reveals patchy ground-glass opacities in pulmonary sarcoidosis resulting from multiple coalescent micronodules with a peribronchovascular and subpleural distribution.

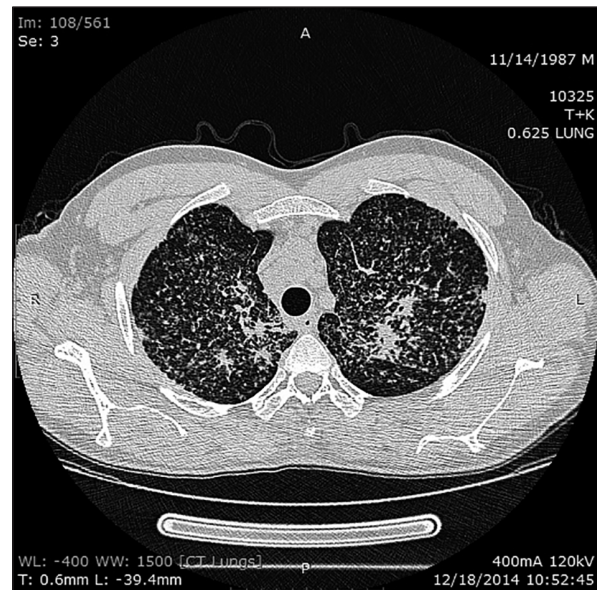


Figure 8. Miliary opacities in sarcoidosis. Axial high-resolution CT scan (pulmonary parenchymal window) shows countless micronodules representing multiple and diffuse granulomas in a random distribution.

OTHER ATYPICAL RADIOLOGIC FEATURES IN SARCOIDOSIS

accumulation of endobronchial granulomas within the bronchial wall, extrinsic compression by enlarged hilar nodes, or distortion of major bronchi by end-stage parenchymal disease, resulting in obstruction and atelectasis^{16, 17}.

Pleural Disease: Pleural sarcoidosis is a rarity (1%–4% of patients) including chylous or hemorrhagic pleural effusion, exudative or transudative pleural effusion, pneumothorax, pleural thickening, or pleural calcification. Pleural plaquelike opacities are most commonly found in sarcoidosis, silicosis, and coal-worker’s pneumoconiosis (figure 10)^{18,19}.

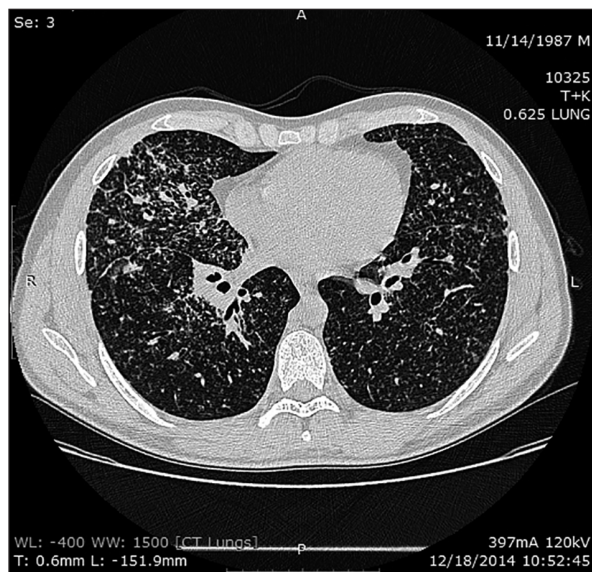


Figure 9. Axial high-resolution CT scan (pulmonary parenchymal window) shows asymmetric subpleural honeycomb-like cysts (medium lobe) and architectural distortion associated with fissure nodularity.



Figure 10. Axial contrast-enhanced CT scan (mediastinal window) in a patient with pulmonary sarcoidosis shows left pleural effusion, diffuse pleural thickening, and bilateral alveolar consolidation.

CONCLUSIONS

Thoracic sarcoidosis manifests with various patterns at radiologic imaging, necessitating an initially broad differential diagnosis that includes lymphoma, tuberculosis, and many other causes of chronic pulmonary infiltrates. The imagistic findings are non-specific or atypical in less than one-half of patients, rarely with no abnormalities seen at thoracic imaging.

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