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THE SPECTRUM OF PULMONARY SARCOIDOSIS PRESENTED ON HRCT- WHAT RADIOLOGIST SHOULD KNOW?- REVIEW ARTICLE

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Abstract

Introduction: Sarcoidosis is a multisystem disease of unknown etiology that very often affects the lung parenchyma with interstitial and granulomatous changes of varying intensity and expression depending on the stage of the disease. Very often mediastinal lymph nodes are affected with changes of different types. It is also called the "great mimicker", because of the various aspects of this enigmatic disease.

In this review we will present typical and atypical variants of pulmonary sarcoidosis according to their distribution detected on HRCT from our experience.

Conclusion: HRCT is the method of choice in the evaluation of pathological changes in pulmonary sarcoidosis. Its capabilities for thin-section imaging and high spatial reconstruction to generate quality images results in better characterization and determination of abnormalities of the lung parenchyma and interstitium.

Key words: sarcoidosis, HRCT, lung, interstitial lung disease

Introduction:

Sarcoidosis is a multisystem disease of unknown etiology that very often affects the lung parenchyma with interstitial and granulomatous changes of varying intensity and expression depending on the stage of the disease.[1] Very often mediastinal lymph nodes are affected with changes of different types. It is also called the great mimicker, because of the various aspects of this enigmatic disease.

Sarcoidosis mainly affects adults over the age of 40 (with a peak in the third decade of life) and has a prevalence of 10-20 cases per 100,000 population. The disease is diffusely spread throughout the world and both sexes are almost equally affected.[2]

Pulmonary involvement is the most common finding (90%) with predominant symptoms of dry cough specially in the morning and dyspnea. Systemic symptoms and symptoms involving other organs are in a significantly smaller percentage (20-30%).[2,3]

The prevalence of the disease is higher among non-smokers than among smokers and is more common in certain occupational groups, such as nurses, firefighters, transport and service workers, although the cause is unknown. Imaging methods such as high-resolution computed tomography (HRCT) play a key role in the diagnosis and monitoring of patients.[4]

HRCT is superior to conventional computed tomography and plain radiogram for the detection and evaluation of subtle parenchymal lesions and abnormalities of lung structures.

It helps us in the prognostic development of the disease and the appropriate treatment. The most important prognostic-predictive factor is the extension of fibrotic changes and reticulation detected on HRCT. HRCT allows us to accurately grade the degrees of the disease with the Scadding score scale.[2]

In the advanced stages of sarcoidosis many different patterns could be seen such as, reticular shadow zones, traction bronchiectasis, architectural distortion of the parenchyma, honeycomb pattern, bullae and paracicatricial emphysema more precisely in the upper and middle lung zones. The lung bases are usually spared. HRCT helps us also in the diagnosis of disease in patients with unusual radiological findings and atypical pulmonary presentations.[5]

HRCT patterns

Typical patterns of lymphadenopathy

The most common pattern is well-defined, bilateral, symmetric hilar and right paratracheal lymph node enlargement (potato nodes) (Figs.1 and 2). Bilateral hilar lymph node enlargement, alone or in combination with mediastinal lymph node enlargement, occurs in an estimated 95% of patients affected with sarcoidosis. Middle mediastinal nodes (at the left paratracheal level, subcarinal level, and level of the aortopulmonary window), prevascular nodes, or both are involved in approximately 50% of patients. Bilateral hilar lymph node enlargement may be a feature of infection (particularly fungal or mycobacterial infection) or malignancy (eg, lymphoma). However, in the absence of specific symptoms or signs, sarcoidosis is the most common cause of bilateral lymph node enlargement.[6]

Other patterns of lymphadenopathy

Occasionally, radiologic findings of lymph node enlargement may be asymmetric or seen in unusual locations. Such findings should lead to the inclusion of entities such as lymphoma or tuberculosis in the differential diagnosis. Isolated unilateral hilar lymph node enlargement (usually on the right side) is seen in less than 5% of case. The enlarged nodes may become calcified. The occurrence of lymph node calcification in sarcoidosis, as in other chronic granulomatous diseases, is closely related to the duration of disease; calcification occurs in 3% of patients after 5 years and in 20% after 10 years. The calcifications may have an amorphous, punctate, popcornlike, or eggshell-like appearance.[7] (Figs.3 and 4)

Typical parenchymal manifestation

Micronodules with perilymphatic distribution

A perilymphatic distribution of micronodular lesions is the most common parenchymal disease pattern seen in patients with pulmonary sarcoidosis (75%–90% of cases). High-resolution CT shows sharply defined, small (1–3 mm in diameter), rounded nodules, usually with a bilateral and symmetric distribution, predominantly but not invariably in the upper and middle zones. The nodules are found most often in the subpleural peribronchovascular interstitium and less often in the interlobular septa (Figs.5 and 6). Extensive peribronchovascular nodularity on HRCT is strongly suggestive of sarcoidosis.[8]

Bilateral perihilar opacities

Confluent nodular opacities that appear on HRCT images as bilateral areas of lung consolidation with irregular edges and blurred margins, radiating from the hilum towards the periphery, are often seen with or without air bronchograms. These areas of consolidation are less homogeneous peripherally and are usually accompanied by micronodules.[9] (Figs.7,8,9)

Fibrotic changes

In most patients, sarcoid granulomas resolve with time. However, in an estimated 20% of patients, fibrosis becomes more prominent over time, producing HRCT and radiographic findings of linear opacities, traction bronchiectasis, and architectural distortion (displacement of fissures and bronchovascular bundles). Fibrosis is seen predominantly in the upper and middle zones, in a patchy distribution.[10]

Atypical pulmonary manifestation

Pulmonary nodules and masses

Pulmonary nodules and masses are seen in 15%–25% of patients with parenchymal opacities.

At HRCT, they usually appear as ill-defined irregular opacities measuring 1–4 cm in diameter that represent coalescent interstitial granulomas (Figs.12,13,14 and 15). These lesions are typically multiple and bilateral, and they may be in perihilar or peripheral regions. Small satellite nodules are often visible at the periphery of these masses, producing an appearance that has been termed the “galaxy sign” (Figs.16 and 17). The galaxy sign was initially described in sarcoidosis but it is not specific for

this condition. It could also be present in active tuberculosis, progressive massive fibrosis or malignancy.

Parenchymal lesions could coalesce, forming conglomerate masses that consist of peribronchovascular fibrous tissue surrounding abnormal conglomerations of perihilar bronchi and vessels. These masses, which are typically seen bilaterally in the upper and middle lung zones, may mimic progressive massive fibrosis.[9,11]

Patchy airspace consolidation and ground glass opacities

Alveolar or airspace opacification is seen in 10%–20% of patients with sarcoidosis.

It is usually bilateral and symmetric and predominantly involves the peribronchovascular regions of the middle and upper zones of the lungs.

Patchy ground-glass opacities are seen in an estimated 40% of patients with parenchymal changes due to pulmonary sarcoidosis. Extensive ground glass opacities are much less common.[12]

Linear opacities and fibrocystic changes

Isolated linear reticular opacities are observed in an estimated 50% of patients with sarcoidosis, but a linear pattern is the predominant radiologic feature of the disease in only 15%–20% of patients. This pattern is produced by interlobular and intralobular septal thickening and usually is seen in the subpleural space in the upper and middle lung zones.

Interlobular septal thickening that is marked and irregular may simulate lymphangitic carcinomatosis. (Figs.18 and 19)

Fibrotic cysts, bullae, honeycombing and paracicatricial emphysema represent advanced-stage sarcoidosis. Fibrotic and cystic lesions typically involve the upper and middle lung zones and follow the large airways in a perihilar distribution.[2,12] (Figs.20 and 21)

Differential diagnosis at imaging

Hilar and mediastinal lymphadenopathy can occur in lymphoma, fungal infection, tuberculosis, and in some cases of either primary lung cancer or metastatic disease to hilar or mediastinal stations.

However, the presence of bilateral hilar and mediastinal lymph node enlargement in the absence of symptoms associated with malignancy or infection, strongly suggests sarcoidosis.

Pneumoconioses such as silicosis can have patterns of calcified lymphadenopathy and perilymphatic nodules like sarcoidosis, including a classic “eggshell” calcification.

Berylliosis has granuloma formation nearly identical to sarcoidosis at pathology. A thorough occupational or environmental exposure history may help distinguish between pneumoconioses and sarcoidosis.[1,6].Consolidation in sarcoidosis can be mistaken for pneumonia and in the chronic setting, can mimic malignancy. In many cases, consolidation is bilateral and relatively symmetric, with a perihilar or peribronchovascular distribution. However, unilateral consolidation, nodules, or reticular opacities can occur in a minority of patients, imitating a wide variety of other conditions. In a minority of cases, honeycombing in sarcoidosis may be lower lobe predominant and may have with an appearance like usual interstitial pneumonia.[8]

Patterns of Lymphadenopathy in Sarcoidosis

Figure 1. **Figure 2.**

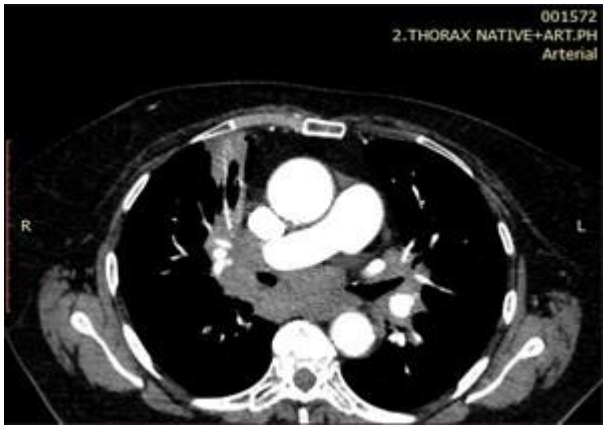


Figure 3.

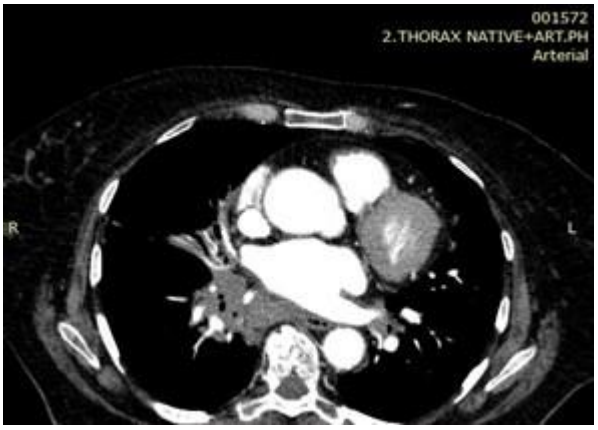
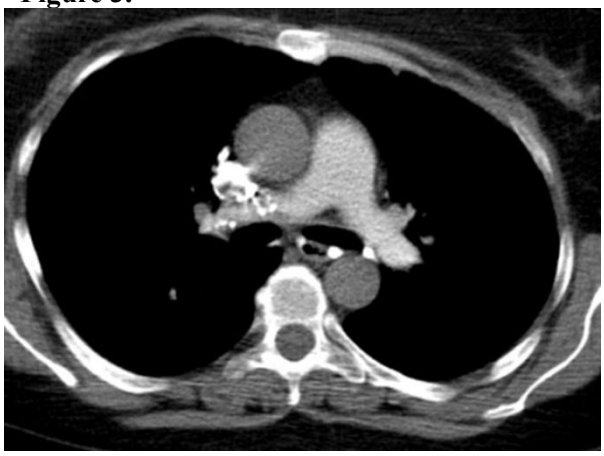
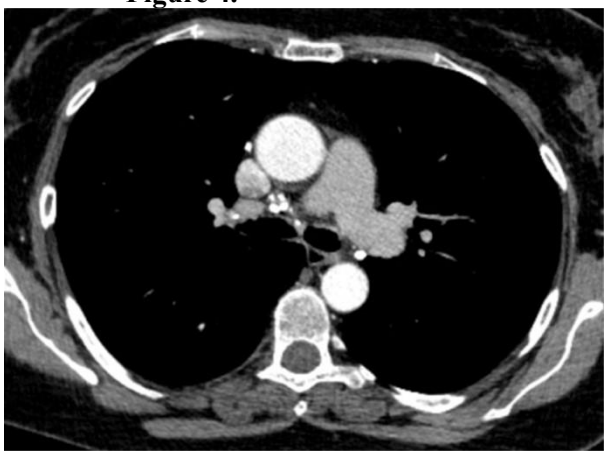


Figure 4.





Typical pattern of sarcoidosis

Figure 5.

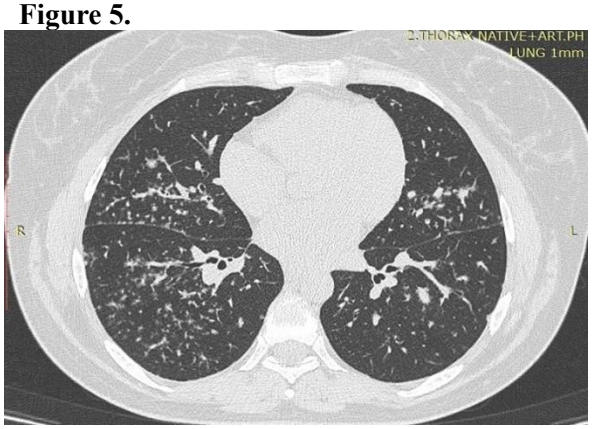
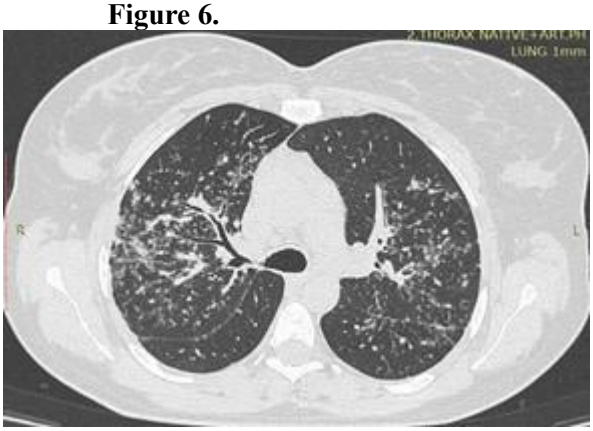


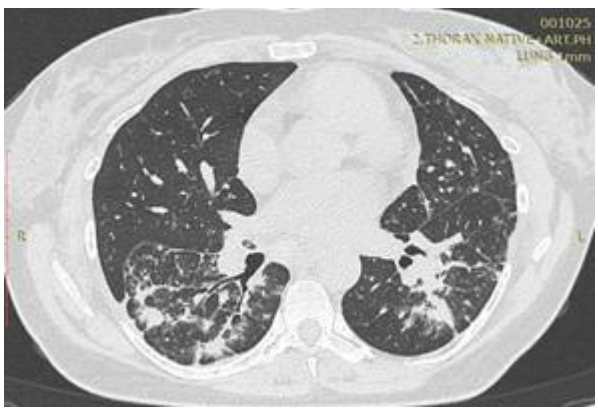
Figure 6.



Bilateral perihilar opacities

Figure 7.

Figure 8.



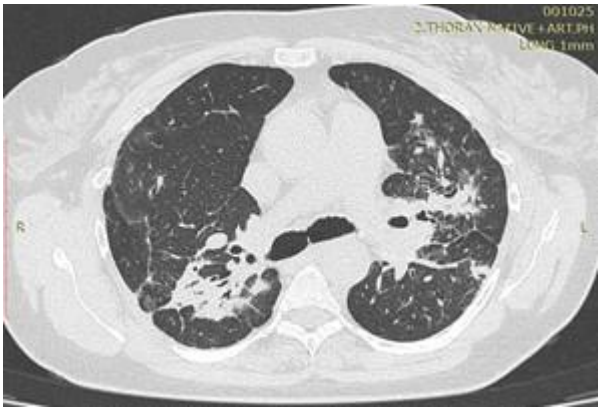
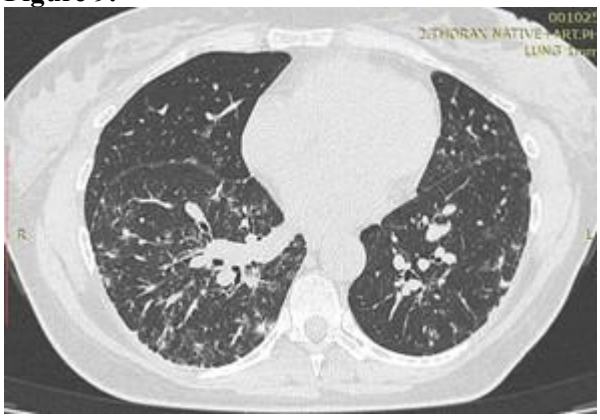
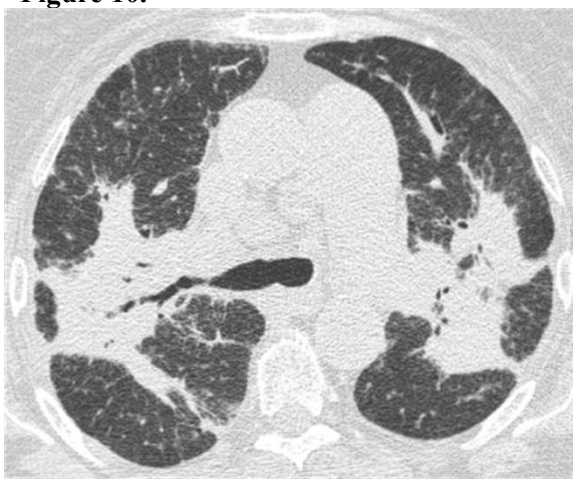


Figure 9.



Fibrotic changes

Figure 10.



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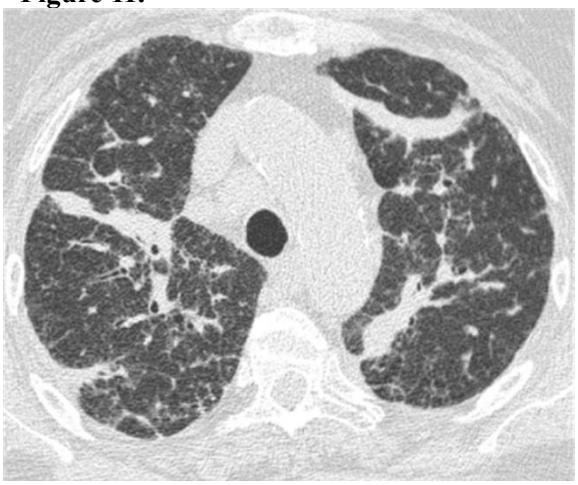


Atypical pulmonary manifestation

Figure 12.

Figure 13.

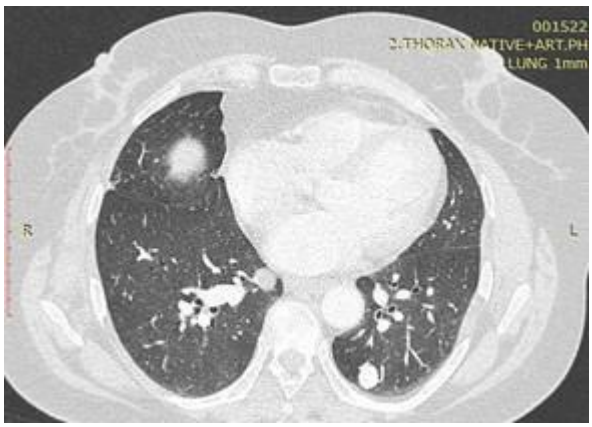


Figure 14.

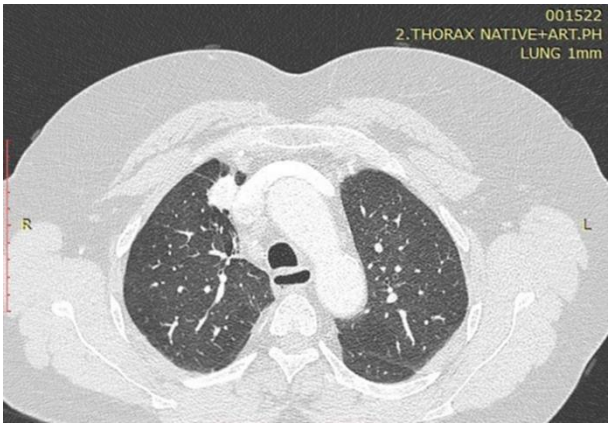
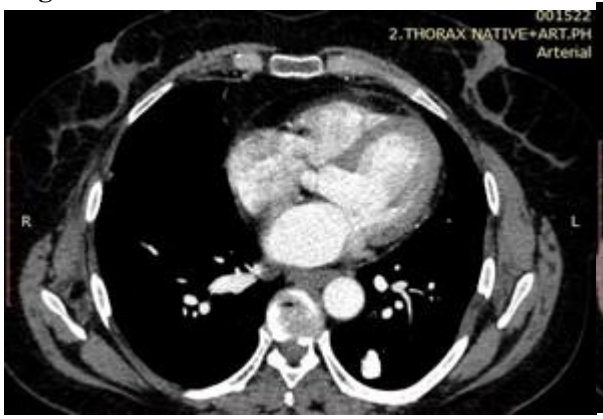
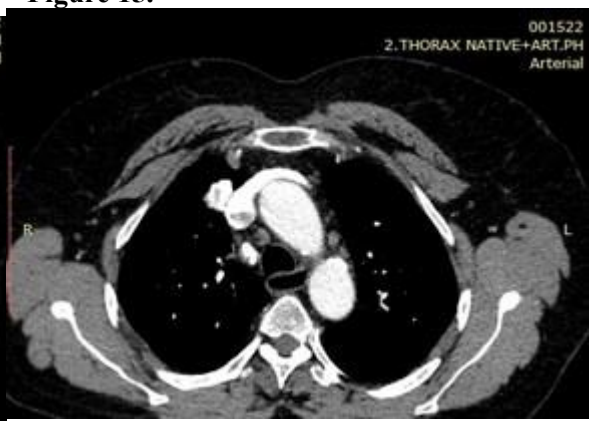


Figure 15.





"Galaxy sign"

Figure 16.

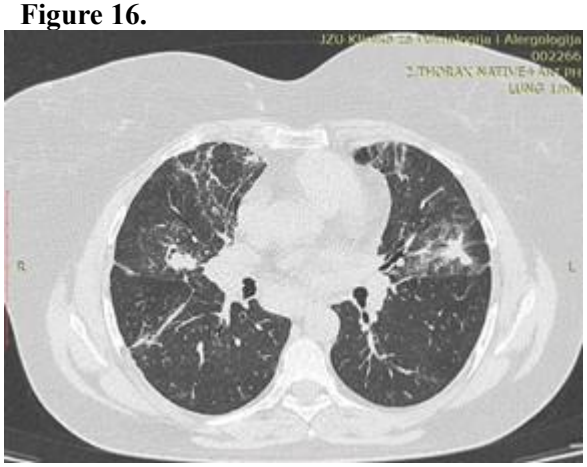
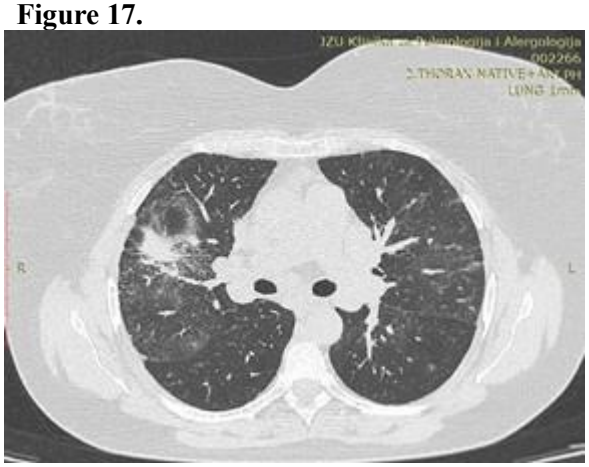


Figure 17.



Linear opacities and fibrocystic changes

Figure 18.

Figure 19.

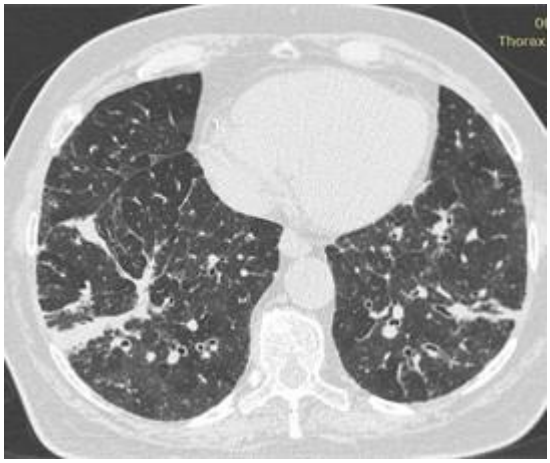


Figure 20.

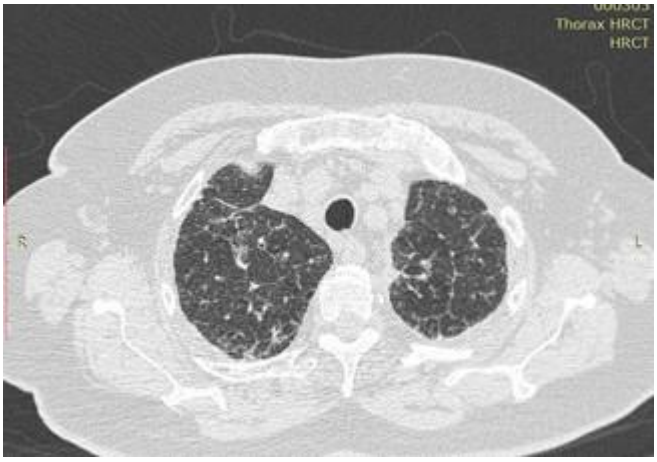
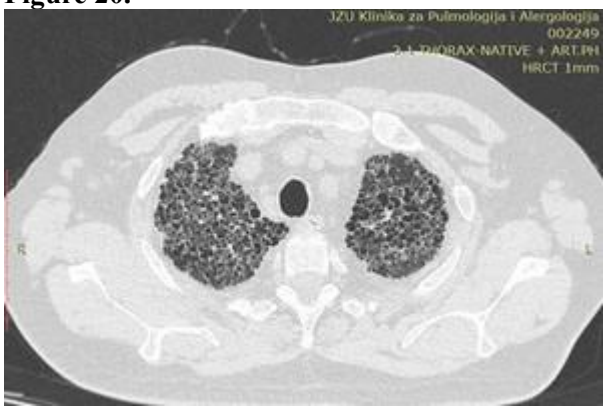
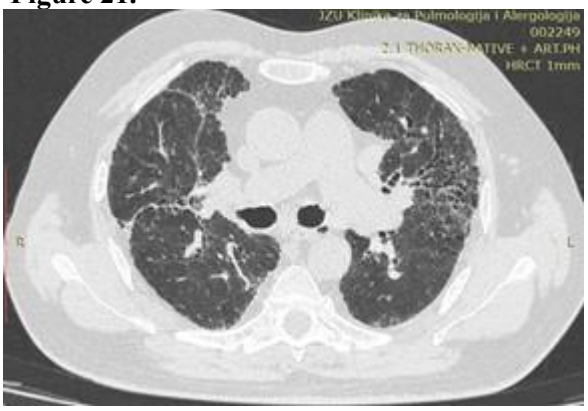


Figure 21.





Discussion:

Sarcoidosis is a multisystemic granulomatous disease of unknown etiology. It mainly affects the thoracic lymph nodes and the lungs.[1]

It is characterized by widespread non- caseating granulomas. There is a wide spectrum of radiologic manifestations in pulmonary sarcoidosis, providing challenges to radiologists. The staging of sarcoidosis, which classifies patients according to their probability of spontaneous remission, is based on the plain chest film findings. Plain chest films are not as sensitive as high resolution computed tomography (HRCT) at detecting involvement of the lymph nodes, lungs, or bronchi. The high resolution CT findings can be typical, practically pathognomic, or atypical. Recognition of the key features of sarcoidosis with knowledge of its pathologic background can often allow for specific diagnosis.[9,13]

Lymphadenopathy is the most common intrathoracic manifestation of sarcoidosis, occurring in the majority of patients at some point in their illness. Symmetric bilateral hilar adenopathy with some form of paratracheal adenopathy is the classic pattern of sarcoidosis and is something that radiologists should always keep in their mind. Symmetry is an important diagnostic feature of hilar adenopathy associated with sarcoidosis because symmetrical adenopathy is unusual in the major diagnostic alternatives including tuberculosis or lymphoma. Faint and amorphous calcification is a common finding in lymphadenopathy due to sarcoidosis. Dense and chunky calcification of the lymph nodes is also commonly seen. Egg-shell calcification, as seen in silicosis, is rare but has been observed. Necrotic or low-density lymph nodes are rarely seen in sarcoidosis.

Presence of small, 1–5 mm nodules with irregular borders is the most common and almost universal finding.[14] These nodules are characteristically in perilymphatic distribution, and therefore,

involve both the bronchovascular bundle and interlobular septa because the lymphatic tract lies along these two anatomic structures in the lung.[14,15,16]

Due to the nodular involvement of the lymphatic system, pulmonary sarcoidosis tends to demonstrate a beaded or irregular appearance with a thickened bronchovascular bundle and interlobular septum, reflecting its granulomatous nature. [17]

The nodular involvement of the lymphatic system is typically symmetrical and demonstrates upper and middle lung zone predominance.

In addition, the presence of architectural distortion associated with nodules is a key finding of pulmonary sarcoidosis (Figs. 4 and 5), which is not seen in other diseases with perilymphatic distribution, including lymphangitic spread of the tumor, pulmonary edema and lymphoma.[13] Multifocal parenchymal opacities of varying sizes occur in 10–20% of patients with pulmonary sarcoidosis. Size of the opacities varies from 1 to 10 cm. The opacities typically have peripheral mid zone predominance and spare costphrenic angle [14].

These opacities represent innumerable coalescent granulomatous lesions on pathology known as “sarcoid galaxy” sign, due to their resemblance to a vast collection of millions and occasionally billions of stars [18] (Fig. 10). Recently, it was reported that “clusters of small nodules”, similar to “sarcoid galaxy sign”, can be seen in pulmonary tuberculosis [19].

Extensive fibrosis occurs in 20–25% of patients with pulmonary sarcoidosis, but takes years to develop. It has a typical distribution of upper and middle zone predominance, and is usually associated with architectural distortion (Fig. 11). Massive parahilar opacities in the upper and upper zones are sometimes seen, simulating progressive massive fibrosis [14,15] (Fig. 12).

Conclusion

HRCT is the method of choice in the evaluation of pathological changes in pulmonary sarcoidosis. Its capabilities for thin-section imaging and high spatial reconstruction to generate quality images result in better characterization and determination of abnormalities of the lung parenchyma and interstitium.

It shows us very precisely the characteristic appearance of nodules and lesions, their distribution and atypical changes and helps us also in staging of sarcoidosis.

HRCT is superior to conventional CT and plain radiography for showing subtle parenchymal lesions and helps us to differentiate active lesions from irreversible fibrosis which is further necessary for appropriate therapy.

Although a variety of high-resolution CT findings are seen in pulmonary sarcoidosis, knowledge of the underlying pathophysiology and recognition of the diagnostic clues of sarcoidosis, including nodular involvement of the lymphatic system with or without bilateral mediastinal and hilar lymphadenopathy, can often allow one to make a specific diagnosis.

References

1. Peeyush KD, Sarvinder S, Megha J, Satyendra NS, Rajat KS. Thoracic sarkoidosis: Imaging with High Resolution Computed tomography, 2017, original article, 1-4.
2. Criado E, Shanchez M, Ramirez J, Arguis P et al. Pulmonary Sarcoidosis: Manifestation of High-Resolution CT with Pathologic Correlation, Chest Imaging, 2010, 1567- 1569
3. Nunes H, Brillet PY, Valeyre D, Brauner MW,Wells AU. Imaging in sarcoidosis. Semin Respir Crit Care Med, 2007; 28: 102-20
4. Keijsers RG, Veltkamp M, Grutters JC. Chest Imaging. Clin Chest Med 2015;36(40) 603-19
5. Webb WR, Higgins CB. Thoracic imaging, 2 nd Edition.philadelphia;Lippincott Williams andWilkins; 2011
6. Silva M, Nunes H, Valeyre D, Sverzellati N.Imaging of sarcoidosis. Clin Rev Allergy Immunol. 2015;49(1): 45-53
7. Wessendorf TE,Bonella F, Costabel U. Diagnosis of Sarcoidosis.Clin Rev Allergy Immunol. 2015;49(1): 54-62
8. Avital M, Halpern IH, Deeb M, Izbicki G. Radiological findings in sarcoidosis. IMAJ. 2008;10: 572-74
9. Ortega IH, Gonzales LL. Update thoracic sarcoidosis. Radiologia. 2011;53(5): 443-48

10. Al Jahdali, Rajiah P, Koteyar SS. Atipical Radiological manifestation of thoracic sarcoidosis: A rewiev and pictorial essay. Annals of Thoracic Medicine. 2013;8(4): 186-96
11. Armengol G, Bernet J, lahaxe L, Levesque H, Marie I. Uncommon manifestation revealing sarcoidosis.Rev Med Interne 2009;30:53-37
12. Martin SG, Kronek LP, Valeyre D, Brauner N, Brillet PY, Nunes H, et al. High-resolution computed tomography to differentiate chronic diffuse interstitial lung diseases with predominant ground-glass pattern using logical analysis of data.eur Radiol.2010;20: 1297- 310.
13. Mizuki Nishinoa, Karen S. Leea, Harumi Itohb, Hiroto Hatabua. The spectrum of pulmonary sarcoidosis: Variations of high-resolution CT findings and clues for specific diagnosis. European Journal of Radiology 73 (2010) 66–73
14. Wilson AG, Hansel DM. Immunologic diseases of the lung. In: Armstrong P, Wilson AG, Dee P, Hansell DM, editors. Imaging of the diseases of the chest. 3rd ed. NW, London: Mosby; 2000. p. 637–88.
15. Webb WR, Muller NL, Naidich DP. High-resolution CT of the lung. 3rd ed Philadelphia: Lippincott Williams &Wilkins; 2000.
16. Itoh H, Nishino M, Hatabu H. Architecture of the lung: morphology and function. J Thorac Imaging 2004;19:221–7.
17. Mimori Y. Sarcoidosis: correlation of HRCT findings with results of pulmonary function tests and serum angiotensin-converting enzyme assay. KurumeMed J 1998;45:247–56.
18. Nakatsu M, Hatabu H, Morikawa K, et al. Large coalescent parenchymal nodules in pulmonary sarcoidosis: “sarcoid galaxy” sign. Am J Roentgenol 2002;178:1389–93.
19. Heo JN, Choi YW, Jeon SC, Park CK. Pulmonary tuberculosis: another disease showing clusters of small nodules. Am J Roentgenol 2005;184:639–42.