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Review

The spectrum of pulmonary sarcoidosis: Variations of high-resolution CT findings and clues for specific diagnosis

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ABSTRACT

Sarcoidosis is a systemic disease of unknown cause, characterized by widespread non-caseating granulomas. There is a wide spectrum of radiologic manifestations in pulmonary sarcoidosis, providing challenges to radiologists. However, recognition of the key features of sarcoidosis with knowledge of its pathologic background can often allow for specific diagnosis. In this review, we describe the variety of high-resolution CT findings in pulmonary sarcoidosis along with its pathologic features as the basis for radiographic manifestations, and discuss the key features on high-resolution CT for the specific diagnosis of pulmonary sarcoidosis.

Contents

1. Introduction

Sarcoidosis is a systemic disease of unknown cause, characterized by widespread non-caseating granulomas. There is a wide spectrum of radiographic manifestations in pulmonary sarcoidosis, which includes any combinations of mediastinal and hilar lymphadenopathy with or without calcification, multifocal parenchymal opacities of varying sizes, diffuse nodular or linear opacities and extensive fibrosis. The variable radiologic manifestations of pulmonary sarcoidosis often provide challenges to radiologists. However, recognition of the key features of pulmonary sarcoidosis coupled with knowledge of the underlying pathology, can often allow one to make the specific diagnosis. In this review, we discuss the variety of high-resolution CT findings in pulmonary sarcoidosis along with the underlying pathology that produces the radiographic manifestations. The key features on high-resolution

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CT for the specific diagnosis of pulmonary sarcoidosis are emphasized.

2. Parenchymal findings

2.1. Diffuse nodular opacities in perilymphatic distribution

Presence of small, 1–5 mm nodules with irregular borders is the most common and almost universal finding [1]. In a review of 44 patients with histologically confirmed sarcoidosis by Brauner et al., nodules were seen in all cases. Nodules were noted as an isolated finding in 19 patients and were associated with other lesions in 25 patients [2].

The nodules seen on high-resolution CT have been shown to correlate with the inflammatory activity of sarcoidosis measured by bronchoalveolar lavage and serum angiotensin-converting enzyme assay [3,4]. Moreover, intra-parenchymal nodules on high-resolution CT were shown to be associated with respiratory functional impairment [5]. 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 [1,6,7]. Of note, the major and minor fissures are the extension of the pleura, and belong to the lymphatic system, and therefore are often involved in pulmonary sarcoidosis. 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 (Figs. 1–3). The strong propensity of sarcoid granulomas to involve the lymphatic system suggests the importance of the lymphatic vessels in the pathogenesis of sarcoidosis [8].

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 (Fig. 6).

Atypical variants of sarcoidosis in the nodular opacities include asymmetric distribution, and lower lung zone predominance (Figs. 7–9).

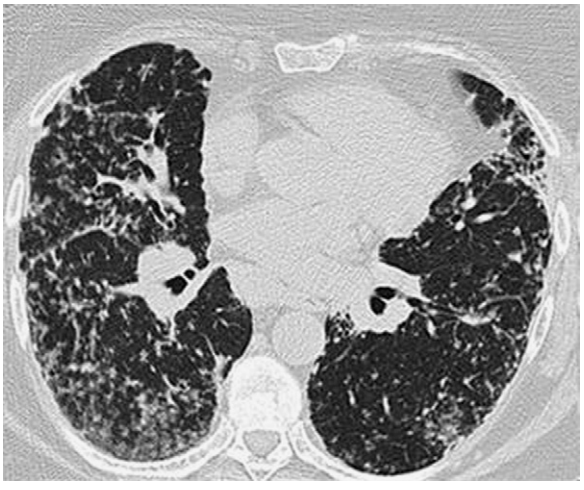

Fig. 1. A 60-year-old man with sarcoidosis. Note the small nodules in perilymphatic distribution, a characteristic high-resolution CT finding of pulmonary sarcoidosis.

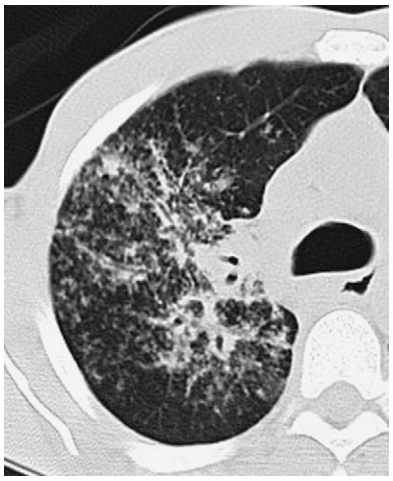

Fig. 2. A 54-year-old man with pulmonary sarcoidosis. Note the beaded appearance with thickened bronchovascular bundle and interlobular septum.

2.2. Multifocal parenchymal opacities of varying sizes

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 [1].

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 [9] (Fig. 10). In the review of histologically confirmed pulmonary sarcoidosis by Nakatsu et al., the sarcoid galaxy was found in 16 (27%) of 59 patients [9]. CT-pathologic correlation revealed that, toward the center of the sarcoid galaxy, granulomas were much more concentrated than at the periphery, giving the appearance of parenchymal opacities rather than small nodules of individual granulomas [9]. Recently, it was reported that “clusters of small nodules”, similar to “sarcoid galaxy sign”, can be seen in pulmonary tuberculosis [10].

2.3. Extensive fibrosis

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 [1,6] (Fig. 12).

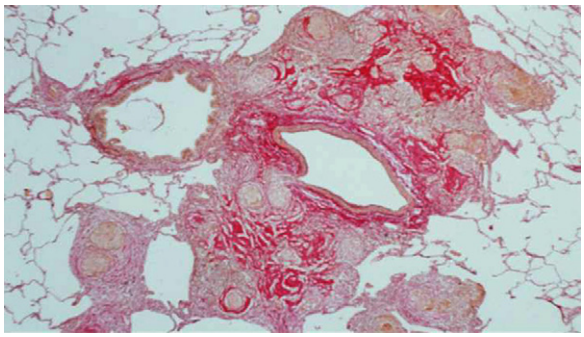

Fig. 3. Microscopic specimen of a lung with sarcoidosis shows the lymphatic pattern, with involvements of the bronchovascular bundle and interlobular septum by non-caseating granulomas.