Extrapulmonary Sarcoidosis: A Chameleon Disease at Imaging

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There is great attention in the literature given to the diagnosis of pulmonary sarcoidosis (SA), where it is established that the combination of factors such as suggestive history, clinical picture, imaging features at computed tomography (CT) such as restrictive disease and lymph nodes enlargement, and (when available) histopathological examination of lung tissues can orient toward the correct diagnosis [1]. In clinical practice, the reality is that the diagnosis of SA is never sure, several disorders can manifest indeed with similar findings [2], and the SA diagnosis can be defined often only “highly probable” when alternative causes are excluded [1, 3]. Also biopsy of the examined tissue can highlight common features of other disorders such as tuberculosis, and sarcoid-like patterns can be found in these patients, going to complicate even more the picture [4-6]. This concept is more true when there is an isolated involvement of extrapulmonary tissues. Organs such as the heart, liver, spleen and the gastroenteric tract can be affected in a not negligible percentage, and clinical manifestations can overlap with those of other diseases (e.g. heart disease with heart dysfunction and myocarditis, liver and spleen with steatosis, splenomegaly and diffusion of parenchymal nodules) and imaging features are not always characteristic to give clues for a presumptive diagnosis [7, 8]. Focal and hypodense lesions can be found as well as non-specific organ enlargement, and these features alone give no elements for a correct diagnosis. These lesions can be easily misdiagnosed with other disorders such as tuberculosis, lymphoma and malignant disease, and the aid of contrast agents can give only a presumptive idea of tissue composition, but not a real estimation of the diagnosis of nature [9-11].

Recently, it has been found that FDG-PET can be useful to demonstrate the inflammatory activity in SA patients. Therefore, a presumptive diagnosis but most of all, a response to the therapy can be detected with the combination of both CT and FDG-PET [12], increasing the likelihood of a correct evaluation before the biopsy [13], in particular for heart disease [14-16].

Advantages are mostly the simultaneous visualization of both PET and CT images, the demonstration of occult sites that can be useful to guide the biopsy, and the view of active extrathoracic sites such as the liver, spleen, and bones, that can be followed-over time to evaluate the correct (or not) response to the therapy [13].

Despite the usefulness of FDG-PET in SA, in particular for the cardiac involvement, there are still some unanswered questions. First, considering the diagnostic potential of FDG-PET, why this examination is not included in the standard workup for SA [17]?

Second, why there are only few studies aimed at investigating the role of FDG-PET in the extracardiac involvement, and most of the evidence is derived only from the reports of singular cases [18]?

Third, an increase could be expected in this diagnostic yield if this technique would be systematically considered in patients with SA?

We believe that this topic should be better investigated and, given the rarity of the disease, prospective multicenter studies could be useful in evaluating and subsequently validating the role of FDG-PET in patients with SA.

More attention should also be given to extrapulmonary manifestations of SA, because these are more insidious than the lung involvement and can lead to significant rates of misdiagnoses and erroneous therapies if are not considered carefully.

Keywords: Sarcoidosis, diagnosis, CT, PET, imaging, lung tissues.

REFERENCES


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