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Case Report

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The "starry night" sign in sarcoidosis

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Introduction

Sarcoidosis is a systemic disease of unknown etiology that can cause organ dysfunction [1-3]. It is most common in young adults between 20 and 50 years of age and in certain races: Swedes, Danes and African-Caribbeans [4-6]. Approximately 4% to 10% of patients have a first-degree relative with sarcoidosis. The disease is diagnosed by protean radiographic, clinical and histopathologic findings [3,7,8]. It is most often defined by the presence of noncaseating granulomatous inflammation that occurs in the lung and thoracic lymph nodes, although different organs may be affected, and the intensity of inflammation often varies throughout the body [9-11]. The etiological agent is unknown. The vast majority of studies suggest that it is the result of a disproportionate immune response in a genetically susceptible individual to certain environmental antigens - bacteria, mycobacterium, viruses, solvents, mold\mildew, pesticides or wood stoves, to name but a few [11-15].

We present an unusual case with a peculiar spleen involvement.

Case report

A previously healthy, non-smoking 31-year- old man, a secondary school teacher, presented at the emergency department with cough, upper left side abdominal pain and the recent appearance of erythema nodosum in the lower limbs. Family history showed the presence of rheumatoid arthritis in the maternal grandmother, and mild asthma in his father. He had no particular hobbies or environmental exposures. He appeared in fair physical condition. Red and painful nodules were present on the anterior surface of both legs. On physical examination, fine crackles could be heard in the lower lungs. The abdomen was meteoric, and the spleen was enlarged and painful on palpation. Body temperature was 37.8°C. The oxygen saturation while he was breathing ambient air was 96%. The complete blood count with differential count was normal. The erythrocyte sedimentation rate was 55 mm per hour (reference range, 0 to 13), and the C-reactive protein level 12.3 mg\L (reference range, <8.0). Anti nuclear antibodies and anti extractable nuclear antigens were negative. Liver function tests were normal. Angiotensin-Converting Enzymes (ACE) had increased to 60 micrograms\L (normal value <40).

Ophthalmological examination was normal. Holter ECG monitoring and echocardiography were negative. An abdominal ultrasound showed the presence of multiple focal hypoechoic lesions in the spleen. A CT scan of the chest showed a diffuse reticular and micronodular involvement of the peribronchovascular interstitium with bilateral enlargement of the hilar, paratracheal and subcarenal lymph nodes. A global spirometry demonstrated a significant reduction in the carbon monoxide diffusion capacity of the lung (60% of predicted value) with normal lung volumes. A total body PET CT scan with F18 FDG (207 MBq Vereos, digital Philips 3D) was performed and the spleen showed intensively avid nodular lesions of various sizes resembling a starry sky pattern (Figure 1) recalling Van Gogh's famous painting "Starry night".

The bronchoalveolar lavage fluid revealed an increase in lymphocytes (33% of total cell count) and an increased CD4/CD8 ratio (5.5). The trans-bronchial biopsy showed the presence of a well-formed and non-necrotizing granuloma in the lung parenchyma and a diagnosis of sarcoidosis was made. The patient was treated with prednisone 40 mg die. Symptoms improved after a couple of weeks. After 8 weeks an abdominal ultrasound revealed a significant reduction in the splenic lesions.

A CT scan, performed after 12 weeks, showed a reduction in interstitial lung involvement and a complete resolution of the splenic lesions. The global spirometry revealed a normalization of diffusion lung capacity (83% of predicted value).

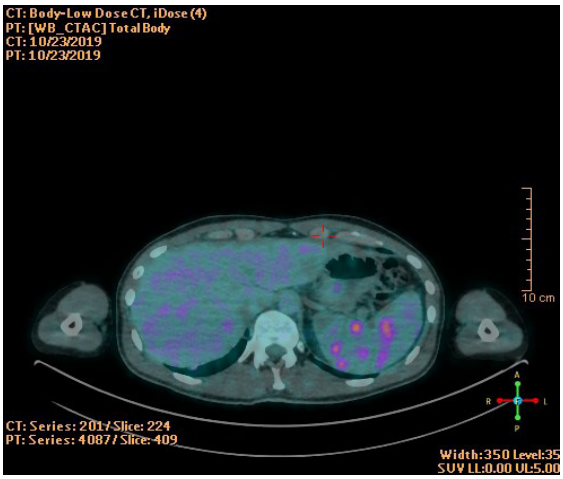

Figure 1:

Discussion

Sarcoidosis should be considered in young and middle aged adults showing dry cough, shortness of breath and constitutional symptoms, such as fever, fatigue or weight loss, particularly in high-prevalence groups such as Blacks and northern European inhabitants [3,11,15].

The differential diagnosis may be a real challenge for the non-specificity of symptoms. A myriad of diseases have similar clinical, radiological and pathological manifestations: infections, above all tuberculosis and histoplasmosis, malignancies, in particular lymphoma, and autoimmune diseases should be excluded in an appropriate clinical setting [2,3,13-15].

Two clinical presentations typical of sarcoidosis are Lofgren and Heerfordt syndrome.

The first is characterized by the acute onset of fever, uveitis and polyarthritis with bilateral hilar adenopathy and erythema nodosum.

The second (uveo parotid fever) is defined by uveitis, fever, parotitis and facial nerve palsy.

Considering extrapulmonary involvement is of paramount importance. In particular, early recognition and treatment of cardiac and neurological sarcoidosis can prevent life-threatening complications. Another target organ is the eye: Anterior or posterior uveitis and optic neuritis can result in rapid and irreversible loss of vision [13-16].

Our patient came to our attention with lung and spleen involvement and erythema nodosum.

In the vast majority of subjects splenic involvement is asymptomatic. The incidence of splenic involvement in sarcoidosis is reported in 6.7 to 77% of patients [16,17]. This high variability depends on the method used for detecting: physical examination, abdominal ultrasound, CT or PET-CT scan. Data relating the role of splenic sarcoidosis on prognosis are sparse and contradictory.

Sarcoid granulomas occur in the white pulp, and can coalesce to produce macroscopically visible nodules.

CT scanning detects sarcoid granulomas in the liver in 50% of cases [16-19].

Clinical signs of hypersplenism: anemia, leukopenia and thrombocytopenia are described [18,19].

FDG PET/CT is not included in the standard workup for sarcoidosis [20,21,22]. However, it may be useful in the evaluation of cardiac, central nervous system and musculoskeletal involvement [20-28]. In some clinical phenotypes it is a useful tool to highlight occult disease areas, disease extent and treatment response, and, above all, to detect the most suitable size for biopsy [29-35,37,37].

Conclusions

In summary, we described the case of a young male presenting with cough, abdominal discomfort and erythema nodosum. Chest CT scan showed a typical pattern of pulmonary sarcoidosis: involvement of peribronchovascular interstitium with bilateral enlargement of the hilar, paratracheal and subcarenal lymph nodes. In order to define disease extent we performed total body PET\CT scan. The definitive diagnosis was reached by trans-bronchial biopsy showing the presence of a well-formed and non-necrotizing granuloma in the lung parenchyma.

We described, for the first time, an interesting PET\CT pattern of spleen involvement in sarcoidosis, and coined a new radiological sign, the "starry night" pattern. Further data are needed to validate this sign from a clinical point of view.

Declarations

Ethics approval and consent to participate: Not applicable.

Human and animal rights: No animals/humans were used for studies that are the basis of this research.

Consent for publication: Not applicable.

Availability of data and materials: Not applicable.

Conflict of interest: The authors declare no conflicts of interest, financial or otherwise.

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