Unusual isolated extrapulmonary sarcoidosis: Case report

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ABSTRACT

Sarcoidosis is a granulomatous condition with unidentified etiology. This disease should be considered in differential diagnosis if non-caseating granulomas is noted in biopsies, because numerous cases presented with no clinical symptoms. Although the lung is the most common organ involved in the disease, there also is an extra-pulmonary form (<10%). It should be noted that the presentation of sarcoidosis may be atypical. In this article, we report a systemic sarcoidosis case, with specific clinical findings: extrathoracic onset with lymph nodes.

1. Introduction

Sarcoidosis is an idiopathic systemic granulomatous disease that has different clinical manifestations based on the organs affected¹. Lung and lymphatic system are the main organs involved². Typical clinical symptoms such as pulmonary symptom are more easily identified.

The typical manifestations of this disease include bilateral hilar lymphadenopathy and pulmonary reticular opacities³. However, up to 30% patients referred with extrapulmonary manifestations, and most common extrapulmonary locations of sarcoidosis are skin, eyes, reticulo-endothelial system and musculoskeletal system⁴. Skin involvement includes different types: papular, nodular, plaque-like, lupus pernio, erythema nodosum and subcutaneous, among which erythema nodosum is best-known⁵. Hepatic sarcoidosis is often asymptomatic, but hepatomegaly (20%) was observed in most cases⁶. Heterogeneity is not observed in images of liver tissue in most cases. However, even in the absence of hepatomegaly in liver biopsy, non-caseating granulomas are observed in about 60% cases. About 15%-35% of sarcoidosis patients showed signs of splenomegaly in images, and hypersplenism may cause anemia, leukopenia, and thrombocytopenia⁷.

Nevertheless, the occurrence of extrapulmonary manifestations in isolated form is rare and only observed in 10% of the cases⁸. Therefore, sarcoidosis patients (especially extrapulmonary) with manifestations of prolonged fever, weight loss, early fatigue, and weakness, are commonly examined under the title fever of unknown origin, and after investigations and differential diagnoses, they are commonly diagnosed with lymphoma or other granulomatous diseases, especially tuberculosis, while other common causes are rejected⁹.

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2. Case report

A 45-year-old man referred with complaints of fever, chills, sweating and weight loss of 10 kg within one month. The patient did not mention any respiratory, digestive, urinary, and neurological symptoms. Hyperthyroidism and inactive chronic hepatitis B (HBS Ag+) could be observed in the patient’s personal history, and there was a family history of diabetes. Besides, he was a smoker and quitted smoking two months before sent to hospital.

Apart from taking thiamazole for hyperthyroidism, the patient did not take any other medicine. The vital signs were stable. In the physical examination, the only positive point was pharyngeal erythema and hyperpigmented lesions on the dorsal surface of his hands, and the patient claimed to have these symptoms four years ago, and no organomegaly or peripheral lymphadenopathy could be touched (Figure 1).

Test results were as follows: Hemoglobin: 11.3 g/dL, HBS Ag+, HBC Ab+, HBe Ag, erythrocyte sedimentation rate: 63 mm/h, alanine aminotransferase: 120 U/L, aspartate aminotransferase: 100 U/L, alkaline phosphatase: 603 U/L, total bilirubin: 0.6 mg/dL, bilirubin (direct): 0.2 mg/dL. Rheumatologic test results were: ANA+, C-ANCA : ¼, PANCA: 6.4, anti-cyclic citrullinated peptide: 1 IU/mL, anti-double stranded DNA (anti-dsDNA): 3 IU/mL, and rheumatoid factor: 30 IU/mL, purified protein derivative skin test: negative, angiotensin-converting enzyme (ACE) level : 35 U/L, and coagulation tests were within the normal range.

Chest x-ray was normal (Figure 2). The abdominal and pelvic ultrasound showed the image of various conglomerate lymph nodes in gastrohepatic and porta hepatis with a maximum diameter of thirty millimeters. The size of the spleen was 131 mm (upper normal limit). For additional investigation, CT scan was done by the medical team. Axial contrast-enhanced CT scan of upper abdomen demonstrates enlarged gastrohepatic and portal lymph nodes, forming a homogeneous conglomerate soft tissue mass in porta hepatis (Figure 3).

In the first place, malignancies and on its top, lymphoma, as well as extrapulmonary tuberculosis, were presented as metastatic or primary liver malignancies. The patient underwent diagnostic laparotomy for excision of the observed masses. The histopathologic results of the masses were lymphoid tissue masses filled with granulomas containing epithelioid cells, a giant cell, and a lymphocyte consistent with non-necrotizing granulomatous lymphadenitis, which indicated sarcoidosis (Figure 4). Tuberculosis rejection by PCR test made the diagnosis of sarcoidosis lymphadenitis the preferred diagnosis.
3. Discussion

The patient was referred with symptoms of fever and weight loss and was examined as a patient with fever of unknown origin. Findings from the patient’s examinations revealed anemia with high sedimentation and high elevations of the transaminases. Then, two isodense masses with liver in porta hepatis were observed in the images. After histological examination, the diagnosis of extrapulmonary sarcoidosis was proposed, which is uncommon in the absence of pulmonary involvement (pulmonary examination for the patient did not prove lung involvement).

In the initial examinations, the patient had hyperpigmented plaque-like lesions on the dorsal surface of his hands. However, due to the fact that the patient had reported their existence for many years before the current disease. Since this type of lesion is not among the most common skin disorders related to the disease, this symptom was neglected. However, considering the fact that skin sarcoidosis involvement in the patient’s clinical presentation is a premature manifestation preceding other manifestations, so a biopsy of the lesions seemed more logical considering the availability of lesions.

Hepatic enzymes increased in the patient’s test results, and considering that the patient had HBS Ag+, the patient seemed to have hepatitis at first glance. However, given the fact that he was monitored regarding hepatitis and he did not have elevated viral load, so there was the possibility of a process other than hepatitis regarding the increase in enzyme levels. The patient’s anemia could by justifies both in the context of chronic disease and in the context of hypersplenism caused by spleen involvement. However, thrombocytopenia and leukopenia were not observed, and the spleen was also reported to be in the maximum normal size in images. Two isodense masses of hepatic hilus indicated lymphadenopathy and involvement of the reticuloendothelial system, while non-necrotizing granulomatous lymphadenitis was also suggested in investigations. In the initial examinations, the ACE of the patient was within normal level, although ACE normally increased by 20%-70% in sarcoidosis patients[10,11]. Also, the patient had hyperthyroidism, which is one of the causes of normal ACE level despite of the presence of sarcoidosis. Treatment for sarcoidosis is not necessary in all cases since the goal is to stop or improve granulomatous inflammation, which underlies the manifestation of the disease that occurs spontaneously in 30%-50% of cases[12]. On the other hand, usual treatment of the disease (glucocorticoids) does not affect the final course of the disease, and relapse is common after the treatment, especially within the first year after the end of treatment[13,14]. There was no pulmonary involvement in our patient and pulmonary involvement with no evidence of liver dysfunction cannot indicate the necessity of treatment, but due to the debilitating manifestations of the disease, fever and weight loss were considerable, thus treatment with prednisolone started in our patient in order to improve symptoms. The patient was discharged after fever was reduced and the symptoms improved. Follow-up was advised.

Conflict of interest statement

The authors report no conflict of interest.

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