

Sarcoidosis onset simulating a unique hepatic metastasis

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Received: 26/02/2018 · **Accepted:** 22/03/2018

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ABSTRACT

Sarcoidosis is a systemic granulomatous disease with an uncertain etiology, characterized by the production of non-necrotizing granulomas. The most frequent presentation is pulmonary and mediastinal, although it might affect any other organ. Hepatic alterations occur in 50 to 65% of the cases. Nevertheless, it is commonly subclinical or detected during a study of the alteration of liver enzymes. It is very unusual that disease onset occurs as an isolated hepatic tumor. A hepatic biopsy is usually required to confirm the diagnosis. A differential diagnosis must be established via any hepatic granulomatous disease, infectious or autoimmune disease as well as the exclusion of malignancy. We present a clinical case of a female diagnosed with an isolated hepatic sarcoidosis that simulated a unique hepatic metastatic lesion. The hepatic biopsy was diagnostic.

Key words: Systemic sarcoidosis. Hepatic sarcoidosis. Granulomatous hepatitis.

INTRODUCTION

Sarcoidosis is a systemic granulomatous disease with an uncertain etiology. The main clinical manifestation is pulmonary or mediastinal. The liver is the third most common organ implicated. Nonetheless, the vast majority of cases appear together with a pulmonary presentation. Only 10% of the patients have an extra-pulmonary isolated disease at sarcoidosis onset as occurred in the present case.

CASE REPORT

The case was a 67-year-old female with previous history of cardiopathy and ischemic stroke who underwent an appendectomy and was in treatment with 300 mg of ASA and 5 mg of bisoprolol. She was referred due to the casual discovery of an unspecific hypoechogenic hepatic lesion via an abdominal echography together with a computed tomography (CT) without contrast. A 20 mm nodule was detected via magnetic resonance imaging (MRI) in the VIII hepatic segment, slightly hypo-intense in the pT1 sequence

and hyperintense in the pT2, suggestive of a metastatic lesion. In addition, a subcentimeter and subpleural nodular lesion was detected at the right paravertebral level, which was observed in a previous study (Fig. 1A). The blood test was normal, as well as the hepatic profile. An increase of the focal metabolism was confirmed in both lesions via positron emission tomography-computed tomography (PET-CT). The capture intensity was compatible with a tumoral process, without other hypercaptured focal points (Fig. 1B). A biopsy of the hepatic lesion was performed. A non-caseific epithelioid granulomas without infection data of acid-alcohol-resistant bacilli or fungi was identified, suggesting hepatic sarcoidosis as a first possibility (Fig. 1C). The patient initiated treatment with 30 mg per day of prednisone. After three months, the subpleural lesion was not visible on a thorax-abdominal CT with contrast and there was also a reduction in size of the hepatic lesion. A stable maintenance of the treatment with 5 mg of prednisone was observed during the last medical exam, six months after the treatment was started.

DISCUSSION

The onset of 90% of the patients with systemic sarcoidosis is characterized by the presence of dyspnea, a non-productive cough and/or mediastinal lymphadenopathies, secondary to the pulmonary and mediastinal manifestations (1). The hepatic manifestation is not uncommon when it is associated with a pulmonary manifestation. Nevertheless, the isolated form is less common.

The infiltration of granulomas from the liver parenchyma usually causes an alteration, generally silent, of the liver enzymes and is responsible for anicteric cholestasis in 10 to 30% of cases. In other cases, jaundice is secondary to

Diéguez-Castillo C, Martín-Lagos-Maldonado A, Ríos-Pelegrina RM, Díaz-Alcázar MM, Roa-Colomo A, Ruiz-Escolano E. Sarcoidosis onset simulating a unique hepatic metastasis. *Rev Esp Enferm Dig* 2018;110(7):466-467.

DOI: 10.17235/reed.2018.5553/2018

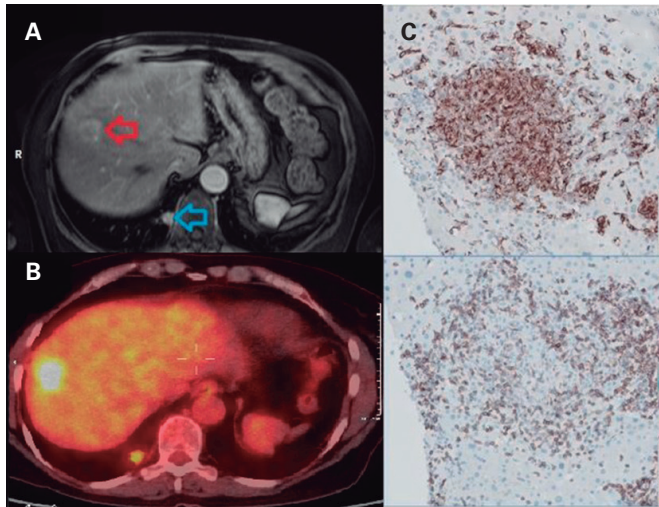


Fig. 1. A. Abdominal magnetic resonance imaging (MRI). A nodular hepatic lesion was identified in the VIII segment, poorly delimited, hypo-intense in the T1 sequence, suggestive of metastasis (red arrow). A non-specific nodular subcentimeter lesion at the right paravertebral level (blue arrow). B. PET-CT. Increased metabolism at the right subpleural level (paraventral) with focal morphology and with mass morphology at the liver parenchyma level (VII segment). C. Histological cut. Immunohistochemical staining with CD68 (top) and CD3 (bottom): non-caseificant granuloma with a central area that consists of aggregates of CD68+ histiocytes and a crown-like structure of surrounding CD3+ T lymphocytes. The double cellular component of sarcoidosis granulomas becomes apparent.

bile obstruction for lymphadenopathies (2). There are other associated symptoms, such as hepatomegaly, splenomegaly, right hipocondric pain, fatigue, fever and/or arthralgia. Only a minority of patients develop hepatic cirrhosis with portal hypertension and/or hepatic failure. Despite this, some patients could develop portal hypertension due to liver infiltration without cirrhosis (3) and exceptionally, Budd-Chiari syndrome (BCS) (4).

Sarcoidosis appearance as a hepatic tumor is unusual and should be used for a differential diagnosis together with other hepatic lesions, such as cholangiocarcinoma or metastatic lesions (5,6). In this case, the presence of a unique hepatic lesion together with a subpleural adenopathy rose a reasonable doubt of a metastatic manifestation.

The absence of specific laboratory or radiologic data complicates the identification of this entity filiation. Therefore,

a diagnosis is established via histological findings. The presence of epithelioid and non-caseificant granulomas in periportal and portal areas with associated macrophages aggregates is diagnostic. Provided that other causes for the development of hepatic granulomas are excluded such as lymphomas (Hodgkin's type), solid tumors, systemic infections (tuberculosis or hepatitis B or C virus), autoimmune diseases (Wegener granulomatosis and primary biliary cholangitis) or drugs.

The treatment of hepatic sarcoidosis is not yet well established. There is controversy about when to initiate treatment and the effect of corticoids. An isolated histologic manifestation does not mean that it is compulsory to start treatment and the asymptomatic patients can be followed up. The majority of authors suggest to start treatment with corticoids, associated or not with ursodeoxycholic acid in symptomatic patients with marked cholestasis and/or high risk for the development of hepatic complications (7). In the present case, the treatment was prescribed due to the suspicion that the subpleural lesion was part of the sarcoidosis manifestations. Immunosuppressors are an alternative when no response is observed and a hepatic transplant is intended for those patients with decompensated hepatic cirrhosis.

In conclusion, this case states that the clinical presentation of sarcoidosis can be complex, making its identification in the presence of isolated extrapulmonary manifestation difficult.

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