© Mattioli 1885

A CASE OF HEPATOSPLENIC SARCOIDOSIS ACCOMPANIED BY HYPERCALCEMIA

Füsun Fakılı¹, Nazan Gülhan Bayram¹, Evren Uzun²

¹Department of Pulmonary Medicine, University of Gaziantep, Gaziantep, Turkey; ²Department of Pathology, University of Gaziantep, Gaziantep, Turkey

ABSTRACT. We present a case diagnosed with hepatosplenic sarcoidosis. Elevated liver enzymes and hypercalcemia were detected in the patient admitted to the hospital with abdominal pain and dyspnea. Abdominal dynamic magnetic resonance imaging showed diffuse, multiple, nodular hyperintense lesions on T2 weighted. Multinuclear giant cells and lymphocytic portal inflammation were seen in the patient's liver biopsy, whose angiotensin-converting enzyme level was high, and hepatosplenic sarcoidosis was diagnosed. Shortness of breath, abdominal pain, and calcium level improved with methylprednisolone treatment.

KEY WORDS: sarcoidosis, extrapulmonary sarcoidosis, hepatosplenomegaly, hypercalcemia

INTRODUCTION

Sarcoidosis is a rare systemic disease with unclear pathogenesis and is seen in different clinics. Lungs and mediastinal lymph nodes are involved in 90% of sarcoidosis cases (1). Extrapulmonary involvement is seen in 30%-50% of patients (2).

A 68-year-old woman presented to the outpatient pulmonary clinic with symptoms of modified Medical Research Council grade II dyspnea, fatigue, and abdominal pain for 3 months. She had hypertension, type 2 diabetes mellitus, and coronary artery disease history. There was no history of smoking or any exposure.

The patient's vital signs were normal and oxygen saturation of 97% on room air. Hepatosplenomegaly was detected in the abdominal examination. Her breath sounds were normal and other systemic examinations were unremarkable. In blood

Correspondence: Füsun Fakılı, MD

laboratory examination, calcium level was 14.6 mg/dL (normal, 8.8-10.6), alanine aminotransferase 54 U/L (normal, 5-35), alkaline phosphatase 617 U/l (normal, 30-120), angiotensin converting enzyme (ACE) level was 243 U/L (normal, 8-52), parathormone level was 2.7 ng/L (normal, 12-88), vitamin D level was 10.8 ug/L (normal, > 30). It was interpreted as secondary hypercalcemia. The blood count was normal and the erythrocyte sedimentation rate was 41mm/h (normal, 0-20). Hepatitis B surface antigen (HBsAg) and anti-HCV were negative.

Abdominal dynamic magnetic resonance imaging (MRI) showed diffuse, multiple, nodular hyperintense lesions on T2-weighted, which did not show enhancement in the liver and spleen parenchyma, and multiple periportal, para-aortocaval lymph nodes. In abdominal MRI, hypointense nodular lesions are seen on T1-weighted images, while on T2-weighted images the nodules are hyperintense compared to the surrounding liver parenchyma (Figure 1).

We decided to perform a biopsy of the lesions in the liver for a definitive diagnosis and to rule out other causes. Mild lymphocytic portal inflammation, ductular proliferation, and portal fibrous expansion were seen in the liver biopsy. Also, multinuclear giant cells were observed in the portal area, and there was no cholestasis, interface hepatitis, or plasma

Received: 2 September 2023

Accepted: 2 October 2023

Department of Pulmonary Medicine, University of Gaziantep, Sahinbey Research Hospital

University Boulevard, 27310, Gaziantep, Turkey

Phone: 090 5322181175

E-mail: fusunfakili@yahoo.com

ORCID: 0000-0002-1602-5240

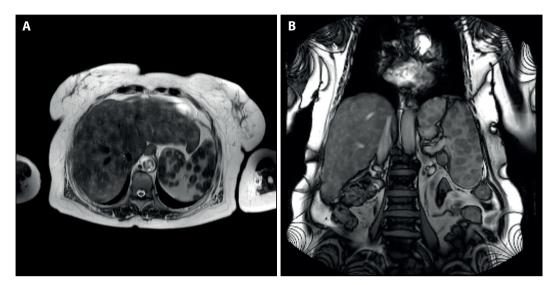


Figure 1. (A) Abdomen MRI. Axial plain. (B) Abdomen MRI. Coronal plain: Diffuse, multiple, nodular hyperintense lesions on T2 weighted, which did not show enhancement in the liver and spleen parenchyma.

cell infiltration in the histopathological liver biopsy analysis. After tuberculosis, fungus, other infectious causes and malignancy were excluded, we diagnosed the patient with hepatosplenic sarcoidosis based on radiological and pathological findings.

The patient diagnosed with hepatosplenic sarcoidosis was started on methylprednisolone 40 mg/day.

The patient's blood calcium level decreased to normal limits after being hydrated. Dyspnea and abdominal pain decreased with steroid. In the first month of treatment, liver enzymes and calcium levels were normal.

The gastrointestinal tract is very rarely the primary site of involvement in sarcoidosis. Most patients with sarcoidosis with hepatic involvement may present with clinical symptoms ranging from asymptomatic to nausea, vomiting, weight loss, pruritus, and abdominal pain. Hepatosplenic sarcoidosis, although rare, can cause cirrhosis, portal hypertension, and liver failure (3). There are no specific laboratory findings for hepatosplenic sarcoidosis. ACE levels have been shown to be higher in patients with sarcoidosis than in those without sarcoidosis (4). T2-weighted MRI are more sensitive to detect non-caseating granulomas as multiple hypodense or T2-hypointense nodules of intermediate size in cases (5). Definitive diagnosis is made by clinical and radiological findings together with the histopathological demonstration of non-necrosing granulomas.

Conflict of Interest: None of the authors have a conflict of interest.

References

- Tana C, Donatiello I, Caputo A, et al. Clinical features, histopathology and differential diagnosis of sarcoidosis. Cells. 2021 Dec 26;11(1):59.
- Sève P, Pacheco Y, Durupt F, et al. Sarcoidosis: a clinical overview from symptoms to diagnosis. Cells. 2021 Mar 31;10(4):766.
- Brugnaro P, Cattelan F, Morelli E, et al. A case of hepatic and bone marrow sarcoidosis with progressive renal failure: diagnostic and therapeutic challenges. Sarcoidosis Vasc Diffuse Lung Dis. 2022 Dec 19;39(4):e2022041.
- 4. Jammal TE, Dhelft F, Pradat P, et al. Diagnostic value of elevated serum angiotensin-converting enzyme and lymphopenia in patients with granulomatous hepatitis. Sarcoidosis Vasc Diffuse Lung Dis. 2023 Sep 13;40(3):e2023031–e2023031.
- Sakai T, Maeda M, Takabatake M, et al. MR imaging of hepatosplenic sarcoidosis. Radiat Med. 1995;13(1):39–41.