Case Report
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Sarcoidosis with hepatic and splenic involvement

Elvin İsazade, MD[1]
Farah Gahramanova, MD[1]
Shabnam Mammadova, MD[1]
Anar Namazov, MD[1]
Nuru Bayramov, MD, PhD, DSc[1]

[1] Department of General Surgery and Transplantology, Azerbaijan Medical University, Baku, Azerbaijan

Abstract: Sarcoidosis is a systemic granulomatous disease of unknown etiology characterized by presence of non-caseating granulomas in the involved organs. The pulmonary interstitium is most commonly affected but extrapulmonary involvement can occur in almost any other organ system. Isolated extrapulmonary involvement has been noted only in around 10% of cases. Here, we describe the case of a 59-year-old patient with isolated hepato-splenic involvement.

Keywords: sarcoidosis, spleen, liver

Correspondence:
Nuru Bayramov, MD, PhD, DSc
Department of General Surgery and Transplantology, Azerbaijan Medical University, Baku, Azerbaijan

E-mail: nurubay2006@yahoo.com
Tel: +994 50 222 2708

Introduction

Sarcoidosis is a multisystem disease characterized by the presence of non-caseating granulomas in affected organs. The pulmonary system is the most common site of involvement, and is affected in about 90% of cases [1, 2]. Sites affected include peripheral lymph nodes (30%), the hepatic system, the spleen, stomach, small bowel, bone, and skin [3]. Bilateral hilar lymphadenopathy is the most common radiological finding. Radiological studies may also show lymphadenopathy, splenomegaly, hepatomegaly, multiple hypointense liver and spleen nodules [4]. However in biopsy and autopsy studies of patients with systemic sarcoidosis, liver involvement was found in about 50–80%, evidence of organ dysfunction is uncommon [5, 6]. Most patients with liver and spleen involvement are asymptomatic. Therefore, the majority of cases are discovered incidentally, frequently by the finding of elevated liver enzymes. Pain in the right upper quadrant of the abdomen, fatigue, pruritus, and jaundice may be associated with liver involvement. Portal hypertension and cirrhosis are complications linked to long-standing hepatic sarcoidosis. Asymptomatic cases do not require treatment. For symptomatic patients, the first line treatment includes corticosteroids or ursodeoxycholic acid. Various immunosuppressant agents can be used as second line agents. Rarely, severe cases require liver transplantation [3].

Case Presentation

A 59-year-old female patient presented to our clinic with a 1-week history of fever, malaise, and fatigue, which began after coronary angiography. She also complained of night sweats, loss of appetite and weight loss (10 kg) in 3 months. She had a medical history of hypertension, type 2 diabetes , and coronary artery disease. Laboratory studies revealed elevated liver enzymes (ALT-62.81 U/L, ALP-524.5 U/L, GGT-301.64 U/L, Total bilirubin-3.98 mg/dL, CRP-36.33 mg/L). Ultrasonography of the abdomen showed hepatosplenomegaly with multiple hypoechoic lesions in the spleen, enlarged lymph nodes at the hepatic and splenic hilum. Chest X-ray showed no significant findings. Abdominal MRI revealed multiple T2 hypointense lesions in the liver and spleen up to 7 mm and 23 mm in size, respectively (lymphoma?); enlarged left paraaortic, interaortocaval, portal lymph nodes up to 12 mm; hepatosplenomegaly; gallbladder sludge (Fig. 1).

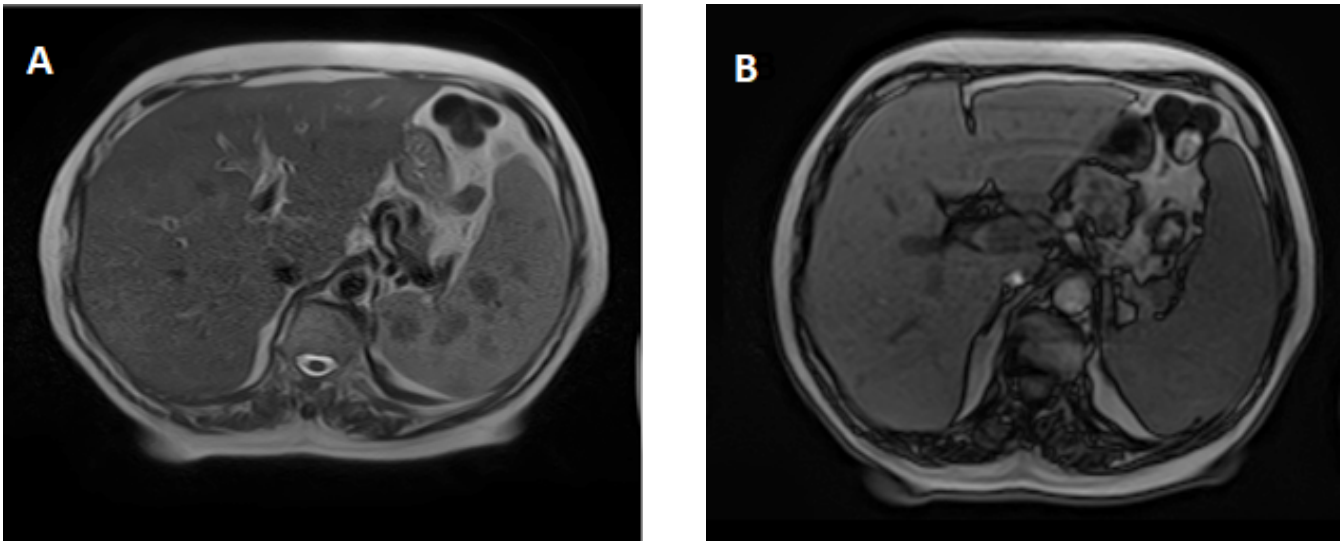

Figure 1. A. Multiple T2 hypointense lesions in the liver and spleen; **B.** Enlarged lymph node at the hepatic hilum.

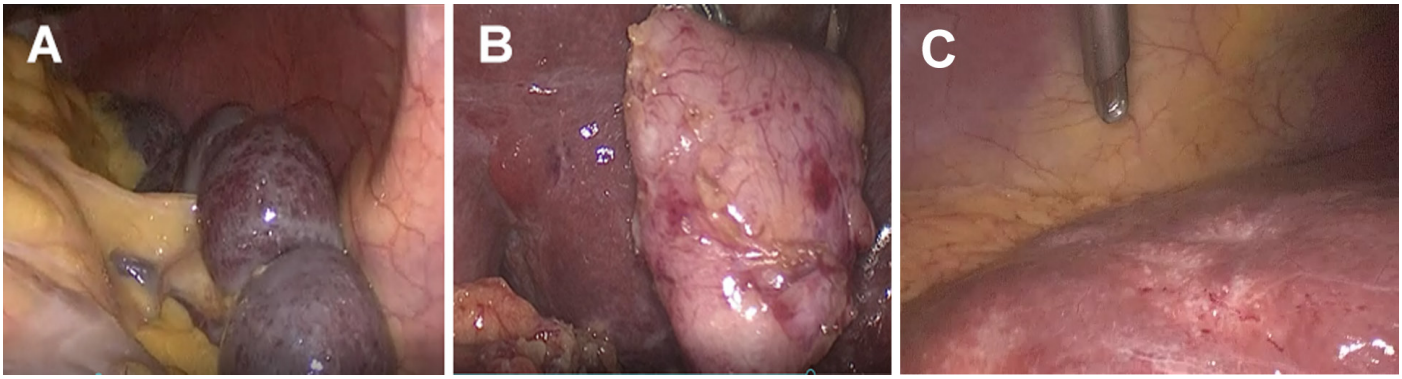

Figure 2. Intraoperative findings: **A.** Multiple nodules throughout spleen; **B.** Enlarged lymph node at the hepatic hilum; **C.** Lesions in the liver.

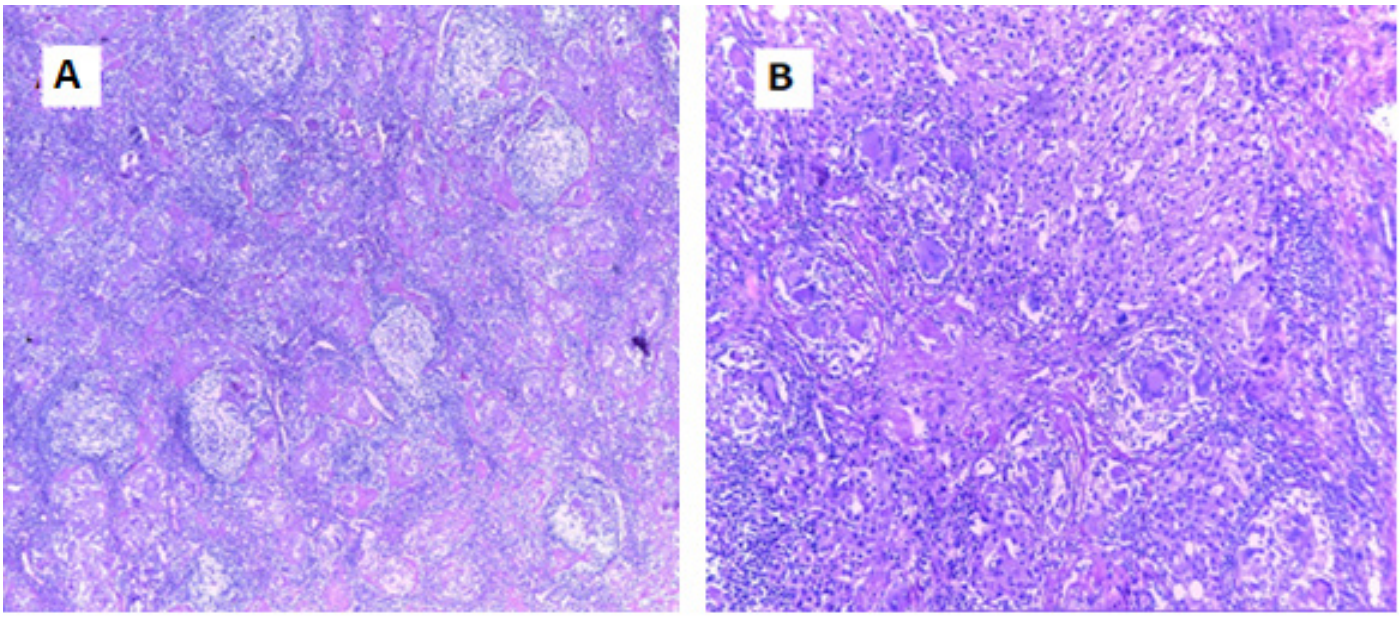

Figure 3. Non-caseating granulomatous inflammation. (H&E) **A.** Lymph node; **B.** Liver tissue

Inital diagnosis was made as lymphoma. Diagnostic laparoscopy was planned to confirm the diagnosis. Under general anesthesia diagnostic laparoscopy, biopsy of liver lesion and hilar lymph node was performed (Fig. 2). Histopathological examination of the biopsy specimens showed non-caseating granulomatous inflammation. Morphological findings suggest sarcoidosis (Fig. 3).

Postoperatively, the patient didn't receive any treatment. She demonstrated spontaneous resolution of her systemic symptoms and normalization of liver enzymes levels over the next 1 month. She has regained her appetite and weight and remained asymptomatic 6 months after diagnosis.

Discussion

Sarcoidosis is a systemic disease which can affect different organs and tissues. It is characterized by the presence of non-caseating granulomas, which can involve multiple organs, in the absence of infections, autoimmune diseases. The prevalence of sarcoidosis is 2–60 per 100,000 people. The most common site of sarcoidosis is the pulmonary system, in more than 90% of cases, patients have pulmonary and mediastinal lymph nodes involvement. Extrapulmonary involvement is rare. Extrapulmonary involvement, in particular in the liver and spleen, is unusual and clinically challenging [1, 2].

Most patients with hepatosplenic sarcoidosis have no symptoms. Abdominal pain, fever, malaise, and weight loss are the most common symptoms in 5-7% of patient. Only 1% of cases, complicated by cirrhosis and portal hypertension, present with ascites and gastroesophageal varice bleeding [3]. As noted above, our patient presented with a complaint fever, fatigue, night sweats, loss of appetite and weight loss.

Laboratory studies are usually non-spesific, dysfunction of liver function tests such as elevated ALT, AST, GGT, and ALP can be observed.

In a minority of patients abnormal findings are revealed on imaging studies. Commonly, USG shows hepatosplenomegaly and enlarged abdominal lymph nodes. Nodular pattern can be detected in low percentage. In these cases, lesions is revealed as small hypoechoic nodules. CT and MRI are more sensitive to detect granulomas. Lesions are detected as hypodense nodules on contrast-enhanced CT. MRI scan reveals T2-hypointense nodules in 5–15% of cases [4, 6]. In our patient, multiple T2 hypointense lesions and enlarged lymph nodes at the hepatic and splenic hilum were detected on MRI. These findings with systemic symptoms raised high suspicion for lymphoma. To establish a specific diagnosis, histopathologic examination was required. We performed laparoscopic biopsy, and the histopathologic examination suggested sarcoidosis.

Pharmacological therapy should be considered for symptomatic patients. Corticosteroids and ursodeoxycholic acid are used as the first line agents. Several immunosuppressants such as azathioprine, methotrexate, cyclosporine have been reported to be effective [3]. About 60% of all symptomatic patients show spontaneous remission as our patient. Asymptomatic patients do not require treatment. Observation is indicated for asymptomatic patients and symptomtic patients who show spontaneus resolution. These patients usually have a good prognosis any medical therapy and remain stable for many years [5, 7].

Conclusion

Sarcoidosis with hepatic and splenic involvement, although rare, should be considered in the differential diagnosis of the patients presenting with systemic symptoms and hepatosplenomegaly with multiple lesions. Histologic examination is necessary for establishing the diagnosis of sarcoidosis. Immunosuppressive therapy should be reserved only for symptomatic patients with sarcoidosis who fails to resolve spontaneously. In our case, we did not give any treatment. The patient demonstrated spontaneous improvement.

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