



Budd-Chiari Syndrome: An Unusual Complication of Sarcoidosis

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Authors' contributions

This work was carried out in collaboration among all authors. Authors Raja Amri and Rabie Ayari designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors MM and ML managed the analyses of the study. Authors HT and MAS managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Hepatic involvement during sarcoidosis is common but rarely symptomatic. Budd-Chiari syndrome is an exceptional but potentially serious complication.

A case of 38-year-old woman was diagnosed with a Budd-Chiari syndrome complicating multisystemic sarcoidosis has been reported in present case study. The patient was treated with hydroxychloroquine and long-term corticosteroid therapy with favorable clinical and radiologic outcomes.

Although BCS is an exceptional complication of hepatic sarcoidosis, it should always be considered. A Doppler ultrasound should be performed in front of any evocating sign to establish an early diagnosis.

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1. INTRODUCTION

Sarcoidosis is an inflammatory disease with poorly known etiologies. It is often caused by an exaggerated immune response to some antigens of susceptible individuals [1].

It is a systemic granulomatosis. The main localization are lymph node and lungs, but it can involve several other organs [2-4]. Extra pulmonary manifestations are reported in half of the cases of systemic sarcoidosis [5]. Liver involvement is often asymptomatic. Clinical involvement is observed in less than 20% of cases. In symptomatic patients, elevated liver enzymes are present in only 10-30% of cases [5,6] and hepatosplenomegaly is noted in about 20% of cases [2].

Bud Chiari syndrome (BCS) is exceptional during sarcoidosis [7].

2. CASE PRESENTATION

In present report, a case of a Tunisian 38-year-old patient with a 4-year history of type 2 diabetes and 8-year history of multisystemic sarcoidosis with skin, lymph node and lung involvement, was reported. The diagnosis was established in front of several findings: disseminated erythematous squamous papular plaques (Fig. 1), hypercalcemia, elevation of angiotensin II converting enzyme, hilar and mediastinal adenopathies with pulmonary parenchymal involvement associated with cutaneous and bronchial granulomatosis.

The patient was treated with hydroxychloroquine and long-term corticosteroid therapy with favorable clinical and radiologic outcomes. During the follow-up, the patient reported right hypochondrial pain. The laboratory workup revealed a prothrombin level of 76%, seven-times-normal anicteric biological cholestasis, and a twice-normal cytolysis. The hepatic angioscanner revealed an absence of opacification of the left and median supra-hepatic veins with perfusion abnormalities in the left liver, all consistent with BCS (Fig. 2). Fibroscopy revealed grade I esophageal varices.

As part of the etiologic evaluation of the BCS, a complete investigation including hereditary and acquired prothrombotic conditions was performed with a negative result.



Fig.1. Extensive erythematous squamous lesions on the back

The diagnosis of primary BCS complicating systemic sarcoidosis was then established. Viral serology and immunological workup were performed in front of a persisting significant cholestasis, but they were negative.

Liver biopsy-puncture revealed gigan-to-cellular granulomatous and epithelioid lesions without necrosis associated with ductopenia and portal fibrosis consistent with sarcoidosis (Fig. 3). The patient was treated with anticoagulant therapy, Prednisolone (40 mg/day) and Ursodeoxycholic Acid (15 mg/Kg/day).

3. DISCUSSION

Liver damage during sarcoidosis is common but rarely symptomatic. In the minority of symptomatic cases, cholestasis is most often discovered during follow-up of known patients with systemic sarcoidosis, which is the case of our patient [8-10]. Liver biology and imaging are recommended during the initial evaluation of sarcoidosis in order to early detect liver damage [11]. Liver biopsy is indicated in cases of moderate or severe disturbances of liver tests [11].



Fig. 2. Coronal section of an abdominal angioscanner showing the absence of opacification of the medial suprahepatic vein (arrow)

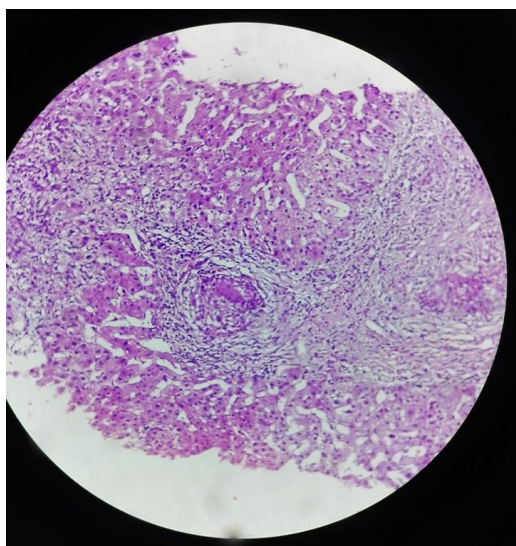


Fig. 3. Hepatic parenchyma containing epithelioid and giantocellular granulomas

Cholestasis appears to be secondary to the progressive destruction of the interlobular bile ducts, due to inflammatory infiltration of the basement membranes and the formation of the portal granuloma. Portal granuloma causes portal tract fibrosis, bile duct destruction and ductopenia [12].

More serious complications such as severe anicteric cholestasis, portal hypertension, BCS or cirrhosis are less common [7,13].

In present case report, an exceptional case of a multisystemic sarcoidosis complicated by a portal cavernoma has been presented. The literature search revealed only 11 cases of BCS related to hepatic sarcoidosis including two pediatric cases [13-23]. The main mechanism of BCS complicating sarcoidosis is not clearly elucidated. Granulomatous lesions are thought to cause external compression of the hepatic veins. They can also directly involve the vascular wall, resulting in lumen narrowing and leading to venous stasis and thrombotic occlusion [12,14,16]. We believe that the development of BCS in our patient may be due to both extrinsic compression and intrinsic granulomatous infiltration of the hepatic veins.

The treatment of hepatic involvement during sarcoidosis is not well codified. Corticosteroid therapy at a dose of 40-60 mg/kg/day is indicated for moderate to severe hepatic impairment, but cannot prevent progression to cirrhosis and portal hypertension [24]. Treatment of BCS associated with sarcoidosis is based on cause management, venous thrombosis prevention and hepatic venous drainage restoration. This may require thrombolytic treatment, angioplasty, transjugular intrahepatic porto-systemic shunt, surgical shunt, or liver transplantation [25-27].

4. CONCLUSION

Although BCS is an exceptional complication of hepatic sarcoidosis, physicians should always consider it. A Doppler ultrasound should be performed in front of any evocating sign, in order to establish an early diagnosis. However, the progression to portal hypertension and hepatocellular failure is unpredictable.

CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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