

Budd-Chiari syndrome: an unusual complication of sarcoidosis.

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.

Keywords: Sarcoidosis, Budd-Chiari syndrome, Cholestasis.

UNDER PEER REVIEW

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 inflammatory disease with main localization in the lungs and lymph nodes, but it can involve several other organs (2-4). Extra pulmonary manifestations are reported in half of the cases of sarcoidosis (5). Liver involvement is often asymptomatic. Clinical manifestations observed in less than 20% of symptomatic patients, are present in only 10-15% of cases (2).

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

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.



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granulomatosis. The lymph node and lungs, several other organs (2-4). Extra pulmonary manifestations are reported in half of the cases of systemic involvement is often 20% of cases. In elevated liver enzymes 30% of cases (5,6) and noted in about 20% of

Fig.1: Extensive squamous lesions

The patient was



erythematous on the back.

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 cytolytic. 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.

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

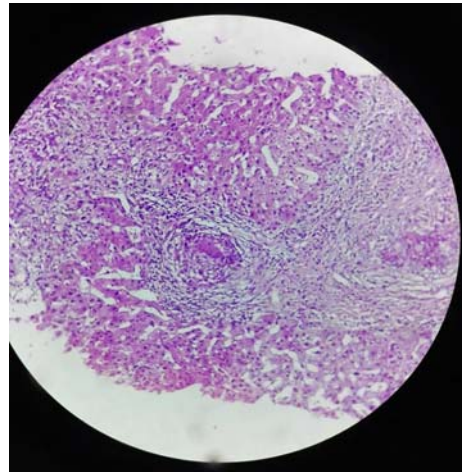
As part of the etiologic evaluation of the BCS, a complete investigation including hereditary and acquired prothrombotic conditions was performed with a negative result.

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 giganto-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 (40mg/day) and Ursodeoxycholic Acid (15mg/Kg/day).

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Fig.3: Hepatic epithelioid and



granulomas.

parenchyma containing
gigantocellular

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).

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).

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.

UNDER PEER REVIEW

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