

Ulcerative Colitis Complicated By Budd- Chiari Syndrome: A Case Report

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Abstract

Case Report

The association of inflammatory bowel disease and thromboembolic events is frequent and secondary to hypercoagulability. Budd–Chiari syndrome has been reported as a rare complication of UC. The incidence of venous thrombosis in UC was found to be 39% in one necropsy study several etiologies are involved, namely coagulopathies and inflammation, the diagnosis is based on imaging (Doppler and abdominal CT). The prognosis depends on the degree of hepatic fibrosis or the interest of an adequate and urgent treatment. We report a case of active ulcerative colitis complicated by hepatic vein thrombosis.

Keywords: chronic inflammatory bowel disease, ulcerative colitis, coagulation, budd-chiari syndrome.

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INTRODUCTION

The Budd–Chiari syndrome (BCS) is a clinical condition caused by hepatic venous outflow obstruction. Ulcerative colitis is associated with increased risk of venous and arterial thromboembolic events. Hepatic vein thrombosis and BCS are a rare extra intestinal complication of UC [1]. A few case reports on hepatic venous involvement with subsequent Budd Chiari syndrome (BCS) are found in the literature, mainly describing adult patients with ulcerative colitis.

In one necropsy study 39% of patients with ulcerative colitis were found on detailed examination to have evidence of venous thrombosis [2]. We report a case of active ulcerative colitis complicated by hepatic vein thrombosis.

OBSERVATION

We report the case of a young patient of 29 years. She has been followed for 5 months for ulcerative colitis treated with 5ASA as a background treatment. She complained of a three weeks history of bloody diarrhea which had recently been increasing in severity. She reported about four to six loose, bloody Stools per day associated with right hypochondrium pain and vomiting. All evolving in a context of asthenia and weight loss.

The medical history was unremarkable, and the patient denied the use of any medication, including

oral contraceptives. On examination, she was clinically anemic and her vital signs were stable. The remainder of the exam was unremarkable. Sigmoidoscopy showed a friable rectal mucosa with contact bleeding and moderate amounts of mucus and blood on the surface.

Investigation revealed Hemoglobin at 7 g/, electrolytes and liver function tests were normal apart from serum albumin 20 g/l. Stool culture was negative as was microscopy for ova, cysts, and parasites. Rectal biopsy showed signs compatible with ulcerative colitis. A barium enema confirmed left sided colitis.

In front of abdominal pain, a doppler ultrasound showed enlarged liver, caudate lobe hypertrophy, the presence of low abundance ascite and no flow in one hepatic vein, with normal flow in portal vein, which was suggestive of BCS. CT scan revealed hepatic vein thrombosis.

The patient was transfused with packed red blood cells and started on oral prednisone. She was also started on heparin intravenous infusion and then transitioned to oral anticoagulant for long-term anticoagulation therapy. She was evaluated for an underlying hypercoagulable state during his illness and did not objectify any abnormality. The evolution was marked by a good clinical and biological improvement.

DISCUSSION

The incidence of inflammatory bowel disease has increased dramatically during the last four decades.

Although IBD primarily affects the bowel, there are many known secondary effects on patients with IBD that are now widely accepted, including an increased risk of venous thromboembolic events [3, 4].

Patients with ulcerative colitis (UC) have an increased risk for both venous and arterial thromboembolism due to hypercoagulability [5, 6]. Budd–Chiari syndrome (BCS) has been reported as a rare complication of UC. The incidence of venous thrombosis in UC was found to be 39% in one necropsy study [2]. But hepatic vein thrombosis and BCS have been reported only as a rare extra intestinal complication of UC. Very few cases occurring in patients without an underlying coagulation disorder have been reported in the literature. BCS is characterized by venous congestion secondary to processes that interrupt or diminish the normal blood flow out of the liver [7].

There are several proposed aetiologies for this increased risk, including thrombocytosis and platelet activation, hyperhomocysteinaemia, increased fibrinogen, impaired fibrinolysis, autoantibodies, elevated procoagulation factors and/or decreased anticoagulation factors and procoagulation mutations, yet no consensus has emerged [8-13].

In general, the cause for a thrombotic tendency in IBD appears to be secondary to a potent prothrombotic stimulus from local and systemic inflammation, and may be related to disease extent and severity [14].

Many studies have shown that thrombosis in IBD is not always related to an underlying genetic or acquired thrombophilia [15, 16].

The intestinal vasculature may be more susceptible to acute arterial events not only because of increased systemic pro-coagulative factors but also because of a localized process that may lead to thrombosis, ischemia, and infarction [17].

BCS can be characterised as a pathophysiological process that results in interruption or diminution of the normal blood flow out of the liver, either within the hepatic veins or inferior vena cava. Symptomatic patients with BCS typically present with ascites, abdominal pain, hepatomegaly and in some cases hepatic necrosis leading to acute liver failure. BCS leads to hepatic congestion, portal hypertension, ascites, oesophageal varices and eventually cirrhosis. It can be classified as acute, subacute or chronic. Furthermore, hepatic venous outflow obstruction in BCS can lead to severe liver failure; without treatment, severe congestion and necrosis lead to fibrosis and ultimately cirrhosis [18, 19].

In some patients, liver function deteriorates so rapidly that they present with acute liver failure. Due to

the various clinical manifestations of BCS, an early histological evaluation to determine severity of the hepatocellular damage is usually recommended, but its role in management and prediction of outcome remains controversial [20]. Histologically, BCS can present with centrilobular congestion, centrilobular necrosis, lobular inflammation, portal inflammation, pericentral fibrosis, periportal fibrosis and/or cirrhosis in the most severe cases.

The diagnostic modalities that have been found to be most helpful are Doppler ultrasound [21] and Computed tomography [22]. Magnetic Resonance Angiography has been shown in a few studies to be more accurate in delineating the hepatic vasculature to more precisely define the location of the obstruction [23]. Nevertheless, clear cut indications for MRI over CT have not been established.

Treatment guidelines for BCS were established in 2009 by the American Association for the Study of Liver Diseases (AASLD). Anticoagulation should be initiated immediately and continued for life unless contraindicated. An extensive workup for secondary causes of hypercoagulability should be performed. In symptomatic patients, percutaneous angiography may be helpful to look for venous obstruction and stents may be placed if necessary. TIPS is reserved for those not improving with anticoagulation and who have failed other management strategies. Liver transplantation should be considered for fulminant liver failure or failure to respond to TIPS. Medical therapy alone is recommended in patients without evidence of ongoing hepatic necrosis [24].

Individuals with IBD after an initial episode of venous thromboembolic events are at increased risk of developing another episode after discontinuation of anticoagulation therapy [25]. Our patient has been kept on anticoagulation therapy and no further episodes of thrombosis have occurred.

BCS is a rare complication of CD and when left untreated can cause significant liver damage and can lead to death. This case recapitulates the need for a high level of suspicion and increased awareness of the venous thromboembolic risks associated with CD. Radiological imaging coupled with histological diagnosis plays an important role in early detection and assessment of the extent of disease in BCS. Appropriate intervention to mitigate hepatic congestion is essential to restore hepatic function and to alleviate portal hypertension.

CONCLUSION

The precise mechanism responsible for increased hypercoagulability in IBD remains unclear. Early recognition and treatment for possible thrombotic complications of CD is critical to prevent potentially fatal events like pulmonary embolism or liver failure.

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