CASE REPORT

Budd-Chiari syndrome in a patient with ulcerative colitis

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SUMMARY

We report a case of 26-year-old man who was admitted on our ward for the evaluation of ascites. He was a known case of inflammatory bowel disease (ulcerative colitis) and was on regular mesalamine therapy. On evaluation, he was having high serum ascites albumin gradient. CT scan of the abdomen revealed features of portal hypertension and non-visualisation of right and middle hepatic veins along with thrombus in inferior vena cava, suggesting a diagnosis of Budd-Chiari syndrome (BCS). There are only few case reports available showing association of BCS with UC. Hence, we are reporting this interesting and rare case.

BACKGROUND

Inflammatory bowel disease (IBD) is a chronic immune-related disorder of the gastrointestinal

Figure 2 Axial contrast-enhanced CT image showing differential enhancement of the liver (dashed arrows) with caudate lobe hypertrophy (asterisk) and gross ascites.

Figure 1 Coronal-reformatted contrast-enhanced CT hepatic venous phase image showing differential enhancement of the liver with thrombosed hepatic vein (black arrow) and gross ascites.

(GI) tract. Disorders of the liver and biliary tract can occur as extraintestinal manifestations of IBD. Budd-Chiari syndrome (BCS) has been described as a rare complication of ulcerative colitis (UC). Patients with UC have a greater tendency to venous thrombosis presenting both clinically and found incidentally at autopsy. The incidence of venous thrombosis in UC was found to be 39% in one autopsy study,¹ but hepatic vein thrombosis in BCS has been reported only as a rare extraintestinal complication of UC.

CASE PRESENTATION

A 26-year-old man was admitted to our hospital with chief complaints of abdominal distension from the last 15 days. There was no history of fever, yellowness of sclera, upper GI bleeding or altered behaviour. There was significant history of UC diagnosed 5 years ago when the patient underwent colonoscopy for chronic bloody diarrhoea. Colonoscopy was suggestive of pancolitis. Diagnosis of ulcerative colitis was confirmed on histopathology. Patient was on regular mesalamine therapy with a history of acute flare-up only once during 5 years. During this presentation, there was no history of worsening of symptoms of UC. There is no history of any indigenous drug intake. Patient was haemodynamically stable and afebrile. There was no icterus or lymphadenopathy. There was no stigma of chronic liver disease. There was temporal loss of fat along with white nails. Per abdomen examination revealed shifting dullness and spleen was

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Table 1 Biochemical and haematological investigations and ascitic fluid analysis

Haemoglobin level	13 g/dL (14–18 g/dL)
Total leucocyte count	5.96×10 ⁹ /L (3.5–12.0×10 ⁹ /L)
Platelets	446×10 ⁹ /L (150–400×10 ⁹ /L)
High sensitivity C reactive protein	25.89 mg/L (<3 mg/L)
Liver function tests	SGPT—31 IU/L (5–35 IU/L), SGOT—30 IU/L (7–40 IU/L) Total bilirubin—0.50 mg/dL (0.2–1.3 mg/dL) Direct bilirubin—0.11 mg/dL (0–0.3 mg/dL) Alkaline Phosphatase(ALP)—248 IU/L (40–150 IU/L)
Serum albumin	22.1 g/L (35–40 g/L)
Serum globulin	27.5 g/L (20–35 g/L)
Kidney function tests	Blood urea—2.33 mmol/L (2.9–8.2 mmol/L) Serum creatinine—33.59 µmol/L (50–110 µmol/L)
Sodium	135 mmol/L (135–145 mmol/L)
Potassium	3.54 mmol/L (3.5–5.1 mmol/L)
Anti-hepatitis C virus rapid test	Non-reactive
Hepatitis B surface antigen rapid test (for hepatitis B)	Negative
HIV rapid test	Non-reactive
Activated partial thromboplastin time	34 s (25–40 s)
Prothrombin time	12.4 s (9–13 s)
International normalised ratio	1.03
Ascitic fluid—microscopic examination	Neubauer chamber count—20 cells/mm ³ Mostly lymphocytes
Serum ascites albumin gradient	1.21

palpable 2 cm below the left costal margin. The patient was further evaluated with biochemical and haematological investigations and ascitic fluid analysis (table 1). Based on investigations and the fact that there was high serum ascites albumin gradient, the patient was further evaluated.

Ultrasonography of the abdomen with venous doppler was done which showed fatty liver with moderate ascites and non-visualisation of right and middle hepatic veins. Further CT abdomen was done which confirmed the findings and showed thrombus in inferior vena cava (IVC) and hepatic veins suggestive of BCS (figures 1–3). Based on clinical and radiological findings, final diagnosis of UC (mild) with BCS was made. Haematological parameters, prothrombin time and activated partial thromboplastin time of the patient were normal. However, extensive coagulation profile of the patient could not be done due to financial constraints. Patient was started on oral anticoagulation

Figure 3 Axial contrast-enhanced CT image showing thickening of sigmoid colon and pseudopolyp formation (arrows) with fat stranding in surrounding region with gross ascites.

therapy (acitrom :composition formula of acenocoumarol)) and mesalamine was continued. The patient was also started on spironolactone, propranolol, and given advice on low salt intake and was discharged. The patient was advised follow-up and international normalised ratio (INR) monitoring.

OUTCOME AND FOLLOW-UP

The patient had come for follow-up after 3 months. The patient is doing well on oral anticoagulants and has improved symptomatically. No untoward side effect of anticoagulants were noticed till now.

DISCUSSION

In BCS, there is interruption or diminution of the normal blood flow out of the liver resulting in venous congestion.² BCS can present in an acute, subacute or chronic form. In an acute form, there is sudden onset of abdominal pain, distention, jaundice, hepatomegaly, ascites and oedema.³ Acute presentation can lead to acute fulminant hepatic failure. The chronic presentations are similar to the acute form but developing much more slowly over the months or years. Also, there is presence of visibly dilated abdominal veins and splenomegaly without encephalopathy.

Hepatic complications of UC include fatty liver, pericholangitis and sclerosing cholangitis, but UC as a precipitant of BCS in the absence of other inheritable causes has been rarely reported. There is increased risk of venous thromboembolism in patients with UC. Moreover, the risk is eight times higher during a flare-up.⁴ There is also increased risk of arterial thromboembolic events.⁵ The incidence of venous thrombosis in UC was found to be 39% in autopsy studies, but hepatic vein or inferior vena cava thrombosis is a rare extraintestinal complication of UC.¹ There are few case reports available suggesting the association between IBD and BCS (table 2).

The exact reason for this increased incidence of thromboembolic events in IBD is, however, not completely understood. No identifiable reason has been found in approximately half of the patients with IBD who develop thromboembolism which reinforces the hypothesis that IBD per se is a risk factor for thrombosis.⁶ There is aberrant behaviour of platelets in both Crohn's disease and UC. Thrombocytosis frequently occurs during the active phase of IBD. There is a high correlation between platelet number and disease activity.⁷ It is likely to be a non-specific response to inflammation; however, the exact reason is still not very well understood. Also, there is lack of sufficient evidence which solidify the association between thrombocytosis and activity of IBD. The platelet count in our patient was normal. This may be due to the fact that the patient was not in an acute flare-up of the UC.

Therapeutic options for BCS (with IBD) are varied and depend on clinical presentation. Anticoagulation should be initiated immediately and continued for life unless contraindicated. An extensive work-up for secondary causes of hypercoagulability should be performed. In symptomatic patients, percutaneous angiography may be done to look for venous obstruction and stents may be placed if necessary. Transjugular intrahepatic portosystemic shunt (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.⁸

Table 2 Review of literature for association between inflammatory bowel disease (IBD) and Budd-Chiari syndrome

Authors, reference no	Patient(s) profile	Remarks
Chesner <i>et al</i> ⁹	54-year-old woman, 35-year-old man	There was association of ulcerative colitis (UC) and fatal Budd-Chiari syndrome (BCS). Progression of the hepatic disorder was unrelated to the activity of the colitis.
Brinson <i>et al</i> ¹⁰	22-year-old woman	Suggested association between active UC and BCS. First reported case in which patient recovered sufficiently to be discharged from the hospital.
Maccini <i>et al</i> ¹¹	35-year-old woman	First reported case of Crohn's disease presenting as BCS
Witteaman <i>et al</i> ¹²	16-year-old woman	Patient with Crohn's disease developed hepatic vein thrombosis possibly due to hypercoagulability during exacerbation of her disease.
Kraut <i>et al</i> ¹³	16-year-old man	Concluded that BCS can occur with IBD even during childhood and non-surgical management is a good option in the short term.
Rahhal <i>et al</i> ¹⁴	14-year-old woman	First reported case of bowel ischaemia secondary to vascular thrombosis in paediatric patients who have IBD.
Park <i>et al</i> ¹⁵	27-year-old woman	There was simultaneous BCS and superior mesenteric vein thrombosis in a patient with UC.
Dacha <i>et al</i> ¹⁶	27-year-old woman	This case highlighted the potential for thrombosis at unusual sites in patients with UC without having classical coagulation abnormalities.
Yilmaz <i>et al</i> ¹⁷	19-year-old man	There was simultaneous presentation of UC and BCS in the patient.

This case, along with the previous reports outlined above, recapitulates the need for a high level of suspicion for BCS in patients with IBD.

Learning points

- Inflammatory bowel disease (IBD) patients can have varied range of intestinal and extraintestinal manifestations.
- Patients with IBD are at increased risk of venous and arterial thrombosis.
- Hepatic vein or inferior vena cava thrombosis is a rare extraintestinal complication of ulcerative colitis.
- There should be a high level of suspicion for Budd-Chiari syndrome in patients with IBD presenting with ascites.

Contributors GKB has produced the report and literature review. VC assisted in the production of the report, the literature review and was also involved in the care of the patient. NM oversaw the creation of the report and was additionally involved in the care of the patient. SB did the radiological investigations of the patient and provided valuable insight into the case.

Competing interests None declared.

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