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Case Report

Budd-Chairi syndrome with intussusception a unique and rare presentation of celiac disease

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ABSTRACT

The association of Budd-Chiari syndrome (BCS) and celiac disease is rare, reported in only 15 cases. None of the case of Budd-Chiari with transient intussusception in celiac disease (in same patient) reported in literature till now. We, hereby, report a 5-year-old male, known case of celiac disease, who presented with pain and distention of abdomen, and failure to thrive for last 2 years. The child had hepatomegaly with fluid thrill. Ultrasonography abdomen showed caudate lobe hypertrophy, transient intussusception with dilated vascular channel and on contrast enhanced computed tomography scan; features suggestive of BCS were found.

Key words: *Budd Chiari syndrome, Celiac disease, Intussusception*

Celiac disease (gluten-sensitive enteropathy) is an immune reaction to wheat gluten and related proteins in genetically predisposed individuals leading to malabsorption, intestinal, and extraintestinal manifestations. It is a malabsorption syndrome showing intestinal villous atrophy, crypt hyperplasia, and drastic reduction in absorptive surface area associated with diarrhea, failure to thrive, vomiting, anorexia, abdominal distension, and foul stool. Affected children are often irritable and unhappy. Many autoimmune conditions such as dermatitis herpetiformis, Type 1 diabetes mellitus, and autoimmune thyroiditis occur in association with celiac sprue [1-3].

Budd-Chiari syndrome (BCS) has been described in celiac disease as an extraintestinal complication as a result of unexplained hypercoagublaty and thrombosis [4]. BCS is a relatively rare condition characterized by an obstruction of the hepatic venous outflow tract and/or a suprahepatic portion of the inferior vena cava (IVC), in the absence of sinusoidal obstruction syndrome, right heart failure or constrictive pericarditis [5]. The association of BCS and celiac disease is very rare and reported in only 15 cases. Except for one, all of these were from North Africa or Southern Europe [4]. None of the case of BCS with transient intussusception in celiac disease has been reported in literature till now. We present such a case in 5-year-old child.

CASE REPORT

A 5-year-old male known case of celiac disease was admitted in Department of Pediatrics with a history of pain and distention of abdomen and failure to thrive for last 2 years. The pain abdomen was generalized, intermittent, and non-radiating. Distention of abdomen was insidious in onset, gradually progressive, intermittent, and temporarily relieved either spontaneously or by some oral medication. The parent also complained that patient not gaining weight and height from past 2 years. He had a history of repeated episodes of diarrhea which was exacerbated by eating wheat products. The patient was diagnosed as a celiac disease 8 months back (by tissue transglutaminase study); since then the patient was on gluten free diet intermittently. Father was also a known case of celiac disease, diagnosed at 11 years of age (Fig. 1).

On general examination, patient had pallor, but no cyanosis, clubbing, and lymphadenopathy. On systemic examination, abdominal inspection shows distended abdomen, everted, and transversally stretched umbilicus and visible dilated veins over the abdomen. On palpation, no tenderness, guarding, or rigidity was present. The patient had hepatomegaly (liver span-11.5 cm) but no spleenomegaly. On percussion, fluid thrill and shifting dullness were present. Other systems were normal on examination.

Routine laboratory studies showed hemoglobin and hematocrit 9.4 g/dl and 32.0%, respectively; white blood cell (WBC) 8340/mm^3, platelet count 433,000/mm^3, mean cell volume 63.6 fL, erythrocyte sedimentation rate 11 mm/h, glucose 93 mg/dl, total bilirubin 0.6 mg/dl, alanine aminotransferase 22 U/L, aspartate aminotransferase 17 U/L, total protein 5.6 g/dl, albumin 3.5 g/dl, Na 141 IU/L, and K 4.1 IU/L. Analysis of ascitic fluid showed total protein 2.8 g/dl, glucose 105 mg/dl, WBC 20/mm^3, with 95% lymphocyte and 5% polymorph. Acid-fast bacillus staining was negative, and Gram staining showed Gram positive cocci but culture of fluid was sterile.

Abdominal USG showed moderate ascites, hepatomegaly (liver - 120 mm) with caudate lobe hypertrophy (4.5 cm×4.6 cm) and altered the liver texture. Hepatic vein lumen was narrow and comma shape collateral in periphery of liver along with

vascular channel at abnormal location in hepatic parenchyma was visualized. An oval hypoechoic mass seen in the right paraumblical region with bright central echo showing targeted configuration. On color Doppler, inner loop showed peristalsis with normal vascularity and showed spontaneous resolution over a period of 1 h suggesting transient intussusception.

Computed tomography scan abdomen showed mild ascites, hepatomegaly with enlarge caudate lobe. Right and left hepatic veins were faintly visualized, and middle hepatic vein was not visualized, along with few venovenal collateral (between portal vein and hepatic vein tributaries) confirming the diagnosis of BCS. However, no obvious portal or hepatic vein or IVC thrombus was identified (Fig. 2).

DISCUSSION

Intussusception is a common cause of bowel obstruction in children and is often idiopathic. In childhood, intussusception usually occurs without obvious cause. Common triggers are Meckel diverticulum, polyps, and tumors, swelling of lymph nodes, cystic fibrosis, and Henoch Schoenlein purpura. Although more common among children, intussusception has not been linked with childhood celiac disease aside from isolated case reports [6]. Despite the intussusception being relatively rare beyond childhood, its association with celiac disease has been described in adults [7-10]. Reported cases of intussusception in celiac disease suggest that it may be asymptomatic, transient, limited to the small intestine and rarely requires surgical intervention and this patient can be managed by strict gluten-free diet [6,11].

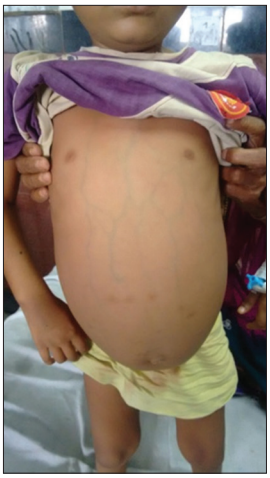

Figure 1: Child with distension of abdomen

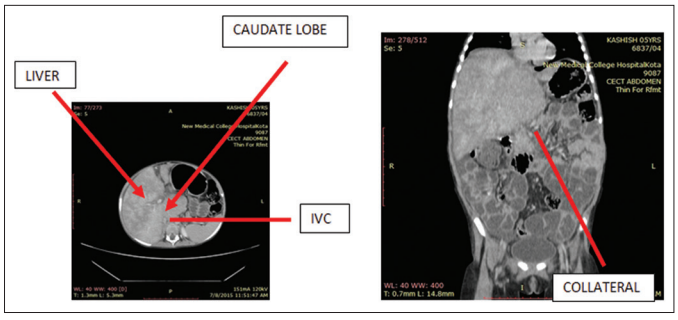

Figure 2: Contrast enhanced CT scan of abdomen showing hepatomegaly with enlarged caudate lobe and veno-venal collaterals

Association between BCS and celiac disease has previously been described in isolated cases in Northern Africa. The appearance of this case suggests that the coexistence of both processes in a single patient is unlikely to be due to environmental or geographical factors, in contrast with previous literature in which this was claimed [1,2]. The unexplained hypercoagulable state in celiac disease may affect the hepatic veins, the mesenteric veins or the splenoportal axis, and causing different syndromes and it is merely the different expression of the same abnormality related to different sites. The reason for the hypercoagulable state may be hyposplenism; various mechanisms have been suggested (1) malabsorption of vitamin K causing protein C, S and antithrombin III deficiency, (2) hyperhomocysteinemia secondary to folic acid deficiency, (3) thrombocytosis, and (4) association with serum lupus anticoagulant [6]. The immunogenetic study showed no association with particular human leukocyte antigens. These patients can be managed conservatively by putting them on strict gluten-free diet [1,12].

CONCLUSION

Although no definitive relationship of celiac disease with Budd-Chiari syndrome and intussusception could be elucidated, these associations must be remembered, especially in the setting of malabsorption. Both the complications of celiac disease are rare and drastic, but they rarely required surgical intervention and can be managed by strict gluten free diet.

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