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Association of inferior vena cava obstruction and Celiac disease in children *(case report)*

Çocuklarda inferior vena kava obstrüksiyonu ve çölyak hastalığı birlikteliği *(olgu sunumu)*

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SUMMARY: *This study describes a 14 years old boy who was diagnosed to have inferior vena cava obstruction and coeliac disease with ultrasonography and intestinal biopsy. Similiar cases are reported in the literature rarely.*

Key Words: **Inferior vena cava obstruction, coeliac disease**

SEVERAL hepatic disorders have been reported in association with coeliac disease (CD). Nonspesific reactive hepatitis, steatosis, and chronic active hepatitis are common and autoimmune liver disease seem to occur more frequently than would be expected from chance alone (1-4). Rare instances of vascular obstructions have also been reported in patients with CD (5-6) but the association of hepatic vein and inferior vena cava obstruction with CD has not yet been fully confirmed (7).

In this report, we present a patient with Budd-Chiari Syndrome due to vena cava inferior obstruction and CD.

CASE REPORT

A 14 year old boy was referred to our department because of hepatosplenomegaly. His weight was 32.5 kg. (normal according to height age) and height was 134 cm (below 3%, sDs:-4.03). Abdominal venous vessels were prominent. The liver was 6 cm and the spleen was 3 cm palpable below the costal margin and palmar erythema was present. His bone age was twelve.

Laboratory tests showed anemia due to iron deficiency. Thrombosit count was normal. Liver enzymes were nearly within normal limits. (AST: 49-35 IU/L, ALT: 28-29 IU/L). Total protein, albumin, bilirubin, gamma glutamyl transferase, alkaline phosphatase, prothrombine time, urea, creatinin and electrolytes were normal.

Ultrasonography showed hepatosplenomegaly, occluded hepatic veins, enlargement of caudate lobe and periportal fibrosis. The inferior vena cava could not be demonstrated suggesting an occlusion. Transfemoral angiography showed total occlusion of the inferior vena cava with a large thrombus (Fig 1). Liver biopsy showed cirrhosis. There was no evidence of hepatic venous congestion or periportal fibrosis. No esophageal varice was seen during endoscopy.

Etiologic investigations considering cirrhosis and thrombus, including; hepatitis B, C, CMV,EBV immunologic profile, ceruloplasmin, serum and urine copper, alpha-1-antitrypsine, sweat chloride test, protein C, S, antithrombine III, acid-ham test were not informative. Endocrinological tests for short stature were normal. Jejunal biopsy showed total villous atrophy, lymphocyte infiltration and crypt hyperplasia. A gluten free diet was institued for probable CD and anticoagulant theraphy was started for thrombosis.

DISCUSSION

In our patient, diagnosis of cirrhosis was estabilished by liver biopsy. No etiological evidence of cirrhosis was found by laboratory investigations. Because of short stature, iron deficiency and cirrhosis of unkown cause, intestinal biopsy was performed for CD suspicion. Histologic evaluation was compatible with CD. CD can be associated with elevated levels of liver enzymes and various degrees of liver injury ranging from reactive hepatitis to chronic active hepatitis, cirrhosis or hepatoma. Mild elevations in aminotransferase levels returning to normal on gluten free diet has been recorded in symptomatic children with CD (1-6). The pathoge-

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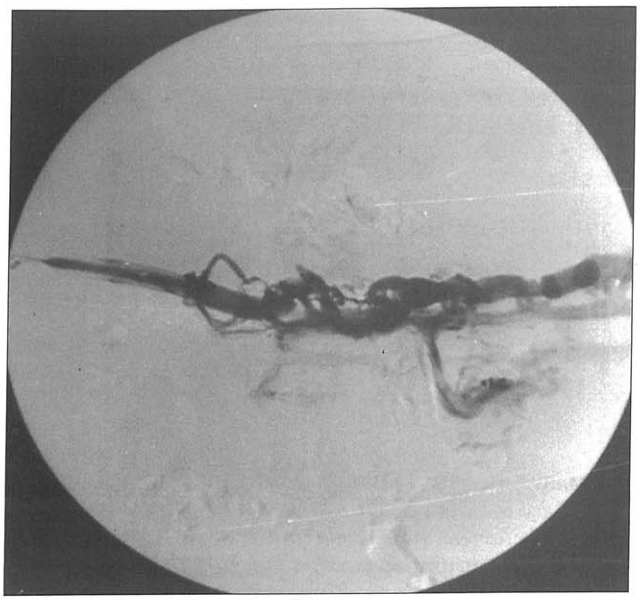

Figure 1. *Transfemoral angiography demonstrating a total occlusion of the inferior vena cava. Note filling of azygos vessels*

nesis of the hypertransaminasemia and liver damage in CD is still unknown. Endogenous or exogenous toxic substances absorbed by the damaged intestinal mucosa might cause liver injury in CD (2).

Rare instances of vascular obstructions, including mesenteric, portal, and hepatic vein have also been reported in patients with CD (5-7). Interestingly, hepatic venous obstruction associated with jejunal atrophy has recently been reported in five North African children and three adult patients (8-10). The cause of hepatic vein obstruction was unclear in most of the eight cases reported to date. In several of these cases, haemostasis disorders were noticed; however these abnormalities are frequent in CD and their role is diffucult to estabilished. In three cases, there was a transient increase in platelet count. This abnormality which might cause thrombosis is frequent and often fluctuating in CD(11-12). In one child antithrombine III level was 60 % of normal values and in two other cases fluctuating deficiency in protein C was noticed. These deficiencies are well known causes of thrombosis. Morover, vitamin K malabsorption due to CD could be responsible for transient protein C or S deficiency since vitamin K is crucial for the activity of these two proteins (13).

The possible causes of hepatic vein obstruction cited above do not explain why there seems to be tendency to have this association in North African patients. Although a random geographic cluster can not be excluded, the role of environmental or genetic factors is more likely. CD and Budd Chiari syndrome are uncommon conditions and it is postulated that this is probably not a change association. It seems likely that the possible cause of caval thrombosis in our patients is CD. On the other hand, we couldn´t find viral, autoimmune and metabolic causes for cirrhosis. Although hepatic venous conjestion or pericentral fibrosis were notfound on histolgic examination, generally, after this pathologic stage of cirrhosis, the differential diagnosis between BCS and other causes cannot be made histologically. Therefore, it is speculated that the possible causes of cirrhosis in this case may be either BCS or CD.

We concluded that further studies are needed for better understanding of the nature of the link between the two entities.

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