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Association Celiac Disease and Budd- Chiari Syndrome: A Case Report: A Rare Association

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Abstract **Case Report**

Budd-Chiari disease associated with celiac disease is a rare phenomenon in medical literature, with the majority of cases reported in the North African region. In this paper, we present the case of a 34-year-old Moroccan woman with Budd-Chiari syndrome associated with celiac disease. It is noteworthy that screening for pro-thrombotic factors yielded negative results, and no other causes besides celiac disease could be identified. This suggests that celiac disease may be a contributing factor in this thrombotic condition.

Keywords: Budd-Chiari syndrome, celiac disease, North Africa, etiology.

INTRODUCTION

Budd-Chiari syndrome covers a range of affections manifested by hepatic venous outflow obstruction, in the absence of right heart failure, constrictive pericarditis or sinusoidal obstruction syndrome. The pathology is relatively uncommon, with an estimated frequency of one case per 100,000 persons [1].

Celiac disease CD is an immune-mediated enteropathy triggered by the ingestion of gluten proteins in genetically susceptible individuals. It is commonly observed in association with a variety of non-gut-related symptoms [2]. Epidemiological studies in North America and Europe have shown that CD is a common disorder, with a prevalence varying between 1/100 and 1/150 [3]. Similarly, studies carried out in North Africa and the Middle East have shown that the prevalence of CD is comparable to that in Western countries [4].

The Budd chiari-celiac disease association is a rare one, first reported in 1990 [5]. Since then, scattered case reports of Budd-Chiari syndrome associated with celiac disease have appeared in the literature. The aim of this case study is to improve our understanding of the epidemiological, clinical and pathogenic features of this disease association.

OBSERVATION

We report the case of Ms E.I, aged 34, treated since the age of 5 for celiac disease, revealed by chronic liquid diarrhoea, a gluten-free diet was instituted and well followed by the patient with a good clinical, biological and endoscopic evolution. She was admitted to our department because of persistent asthenia for 6 years, with no associated digestive or non- digestive symptoms. The patient's general examination was hemodynamically and respiratorily stable. Clinical examination revealed an undistended abdomen, with epigastric collateral venous circulation. Splenomegaly was present, with no hepatomegaly, or mass, and no other abnormalities.

Biological tests revealed pancytopenia, with anemia (hemoglobin 11), leukopenia (3180) associated with lymphopenia (369) and thrombocytopenia (76000). In addition, a slight nutritional deficit was noted, with hypoalbuminemia at 30 g/l and hypocholesterolemia at 1.18 g/l. Prothrombin time was 92%, and there were no abnormal liver enzymes.

Abdominal ultrasound, supplemented by a Doppler study, showed a dysmorphic liver with atrophy of the right lateral sector and segment IV. The suprahepatic veins were small, with an echogenic cord at the level of the median Suprahepatic Vein, and the portal trunk was permeable. There was also significant dilatation of the spleno-mesaraic trunk and splenic vein,

with collateral circulation in the splenic hilum. The diagnosis of Budd-Chiari syndrome was suspected. An abdominal angioscanner was then carried out, showing atrophy of the right lateral sector and segment IV, and a small aspect of the hepatic veins which were not opacified during venous time. This suggested a diagnosis of Budd-Chiari syndrome associated with celiac disease in remission.

Therefore, as part of the search for signs of portal hypertension, an oeso-gastroduodenal fibroscopy was performed, revealing grade II Esophaeal Varices with no signs of active bleeding, and a slight effacement of the duodenal folds. However, histological examination of duodenal biopsies was normal, and tests for celiac disease antibodies (anti-transglutaminase IgA antibodies) were negative, indicating good compliance with and a positive response to the gluten-free diet.

However, as part of the etiological work-up for his Budd Chiari;

Myelogram and bone marrow cell culture were within normal limits. However, it was not possible to perform the JAK2 mutation due to resource limitations. Thrombophilia test, including homocysteinemia, antithrombin, protein S, protein C, antiphospholipid antibodies and factor V Leiden mutation, were negative. It is therefore an idiopathic Budd-Chiari syndrome requiring close monitoring in the day hospital.

DISCUSSION

The Budd Chiari syndrome is a group of clinical manifestations resulting from an obstruction to hepatic venous drainage. The obstruction may be located in the suprahepatic veins, or in the suprahepatic inferior vena cava, up to its junction with the right atrium [6]. The result is sinusoid congestion, hepatocyte ischemia and portal hypertension. [7].

Other common causes of Budd-Chiari syndrome include coagulation disorders such as antiphospholipid antibody syndrome, isolated deficiencies of protein C, protein S or antithrombin, as well as heterozygous or homozygous mutations of factor V Leiden or the prothrombin gene, and complex thrombophilias [9, 10]. Other rarer causes, such as paroxysmal nocturnal hemoglobinuria, should also be investigated. However, the etiology of Budd-Chiari syndrome remains undetermined despite an exhaustive work-up in over 10% of cases.

In the case of our patient, we did not find any of these factors responsible for thrombosis. However, the study of the JAK2 mutation could not be carried out (due to financial constraints).

Studies have shown that various extra-intestinal disorders have been associated with Celiac Disease, including diabetes type 1, dermatitis herpetiformis and autoimmune thyroiditis. Other hepatobiliary disorders such as mild isolated hypertransaminasemia, autoimmune hepatitis, primary biliary cirrhosis and primary sclerosing cholangitis have also been reported [11].

Budd-Chiari syndrome associated with CD is more prevalent in North African countries, perhaps due to the high prevalence of celiac disease in North Africa [12], suggesting that the association may be fortuitous. A case series showed that the majority of reported patients belonged to the female sex and presented in the third or fourth decade [13]. Similarly, our patient was also female in her third decade of life.

In 61% of cases of Budd chiari syndrome associated with celiac disease, no specific thrombotic etiology has been found, as in our case. Conversely, an underlying thrombotic condition can be detected in over 80% of patients with isolated Budd-Chiari syndrome [14]. It is therefore possible that celiac disease may play a role in the thrombotic process leading to Budd-Chiari syndrome.

It is possible that the hyposplenism commonly found in patients with celiac disease may, through thrombocytosis, provide the basis for thrombosis; however, other mechanisms have been suggested to explain the pro-thrombotic potential of celiac disease, including vitamin K malabsorption causing protein C, S and antithrombin III deficiency, hyperhomocysteinemia secondary to folate deficiency [15].

The effect of a gluten-free diet on the evolution of Budd-Chiari syndrome in celiac disease has not been frequently reported [16]. In our patient, Budd chiari syndrome had developed despite strict adherence to a gluten-free diet, as shown by the histopathological examination, which was normal, and the negative celiac serology.

In a study conducted in Algeria, the authors evaluated the etiology of Budd chiari syndrome in 116 patients [17]. In this study, celiac disease was found in 11.4% of patients, and only 40% of these patients had an underlying thrombophilia associated with celiac disease. On the basis of their findings, the authors proposed a systematic search for celiac disease in the etiological work-up of Budd Chiari Syndrome.

CONCLUSION

Budd-Chiari syndrome associated with celiac disease is a rare phenomenon in the medical literature, with an annual incidence of less than five per million. The majority of cases are reported in the North African region [18].

Further studies are needed to understand the pathogenesis of this association, which is probably not a coincidental one, although no link between these diseases and the ethnic origin of the subjects has been

demonstrated. The frequency of celiac disease justifies its consideration when Budd Chiari syndrome is diagnosed in our region.

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