Occult celiac disease presenting as splenic vein thrombosis

Celiac disease may present with varying symptoms of malabsorption and nutrient deficiency. Venous-thromboembolism is an uncommon presentation of celiac disease. We present a case of splenic vein thrombosis that was later on found to have celiac disease.

A 39-year-old lady was transferred from another hospital with complaints of moderate to severe left upper quadrant constant pain with radiation to the left shoulder since 7 days. On examination, there was tachycardia and tenderness on palpation in the left upper quadrant. The blood investigations revealed Hb 11 gm/dL, total leukocyte count 13,400/mm³, platelet count 1.7×10⁹/mm³. Liver and kidney biochemical tests, and amylase and lipase were normal. Chest x-ray revealed raised left dome of diaphragm. Ultrasound of the abdomen revealed thrombosis of the splenic vein and large hypoechoic density in the spleen suggestive of splenic infarct. These findings were confirmed by CECT. Blood was sent for procoagulant factors and patient was started on low molecular weight heparin and antibiotics. The pain gradually resolved. The leukocyte count returned to normal and patient was discharged on oral anticoagulants. The serum homocysteine levels were high (20 µmol/L, normal value 5-15). Protein C and S, anti-thrombin III levels were normal. ANA, DsDNA and lupus anticoagulant were not detected. Folic acid was added to the treatment.

The patient continued to do well for 6 weeks when she developed diarrhea, and anorexia. There was transient improvement with antibiotics. At UGI endoscopy, the duodenal mucosa appeared to be flattened; duodenal biopsy showed partial villous atrophy with increased intraepithelial lymphocytes. Ig A antibodies against tissue transglutaminase were elevated. She was thus diagnosed as celiac disease. The patient improved on gluten-free diet. At follow up, there was partial recanalization of splenic vein with collaterals formation around it.

The possible association of celiac disease and venous thrombosis was first reported in 1970.¹ Subsequently, thrombosis at other sites including hepatic veins,²,³ portal vein, cavernous sinus,⁴ and lower limb veins⁵ has been reported in isolated case reports. It is unlikely that these are chance observations. One report documented deficiency of protein S in the patient.⁴ Our patient had splenic vein thrombosis associated with celiac disease. Interestingly, unlike in previous reports, celiac disease in our patient manifested after the thrombotic phenomenon.

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