Gutfile

Coeliac disease presenting as variceal haemorrhage

CASE REPORT

A 59 year old woman was referred for investigation of lethargy, ankle oedema, and an elevated alkaline phosphatase. Sixteen years previously her entire jejunum had been resected at another hospital for acute mesenteric vein thrombosis.

On investigation, alkaline phosphatase and prothrombin time were raised, and calcium, haemoglobin, and red cell folate were low. She was heterozygous for the prothrombin 20210A gene mutation and table 1 summarises the results of her thrombophilia screen. Activated protein C resistance, anti-thrombin activity, anticardiolipin antibodies, haemolysis, and paroxysmal nocturnal haemoglobinuria screen were normal or negative. Abdominal ultrasonography revealed splenomegaly and cavernous transformation of the portal vein with numerous venous collaterals.

While undergoing these investigations she experienced melaena. Urgent endoscopy revealed two columns of oesophageal varices with stigmata of recent haemorrhage which were eradicated by variceal band ligation. An incidental finding of scalloping of distal duodenum mucosal folds was observed and histology revealed subtotal villous atrophy consistent with coeliac disease. Subsequent IgA antiendomysial antibody was positive. Review of previously resected jejunum confirmed the changes of venous gangrene, in addition to subtotal

villous atrophy at the proximal resection margin.

Gluten restriction resulted in 8 kg weight gain, normalisation of folate, calcium, alkaline phosphatase, and coagulation screen, and a dramatic improvement in duodenal histology. Thrombophilia screen was repeated (table 1). With heterozygosity for the prothrombin 20210A gene mutation, the only persisting prothrombotic abnormality, long term anticoagulation, was avoided.

DISCUSSION

A prothrombotic disorder is identified in approximately 70% of patients with portal or mesenteric venous thrombosis.^{1 2} In most cases several acquired or inherited prothrombotic factors coexist.^{1 3}

Although intra-abdominal venous thrombosis complicating coeliac disease has been reported, coeliac disease presenting with variceal haemorrhage secondary to portal vein thrombosis has not been described. Our patient was heterozygous for the prothrombin 20210A gene mutation, which is found in 2.3% of healthy controls⁴ and hence it is likely that a "second hit" is necessary to stimulate thrombogenesis. In our patient, this was probably a reversible reduction in vitamin K dependant protein C and S levels due to coeliac disease. In addition, both enteric protein loss with net jejunal secretion of water and

hyperhomocysteinaemia secondary to folate deficiency may contribute to a reversible prothrombotic milieu in coeliac disease.

Anticoagulation is indicated in patients with acute mesenteric vein thrombosis⁵ but long term anticoagulation in patients presenting with chronic portal thrombosis is only indicated if there is a persistent prothrombotic risk. Although our patient remains heterozygous for the prothrombin 20210A gene mutation, gluten restriction corrected all acquired prothrombotic abnormalities which allowed her to avoid long term anticoagulation with its associated risks.

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Table 1 Levels of vitamin K dependant procoagulant and anticoagulant factors prior to and following six months of a gluten free diet

	At presentation	After six months of gluten restriction
Protein C activity (IU/dl) (normal range 74-128)	50	74
Total protein S antigen (IU/dl) (normal range 64-166)	43	81
Free protein S antigen (IU/dl) (normal range 51-140)	42	66
Factor VII (IU/dl) (normal range 85-251)	36	89
Factor IXc (IU/dl) (normal range 56-131)	50	80
Factor Xc (IU/dl) (normal range 54-123)	52	69