Coeliac disease presenting as acute bleeding disorders

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Summary
Two young women presented with acute bleeding disorders and were subsequently found to have coeliac disease.

Introduction
The presentation of coeliac disease as an acute bleeding disorder is an unusual event. Two recent cases are reported in which the bleeding disorder was of such severity that the patients presented as emergencies to haematologists.

Case 1
A previously well, 27-year-old woman was admitted to an acute haematology unit with multiple severe bruising. For 3 months before admission she had been having vague colicky abdominal pain with pale, bulky, offensive stools which were difficult to flush away. She had lost 9.5 kg in weight. Two weeks before admission she had developed spontaneous bruising of her limbs. She had no melaena or haematuria. She was unemployed, a non-smoker, and she drank three pints of beer nightly. She had a normal diet and was on no drugs.

Examination showed her to be pale with multiple old and fresh bruises as well as large haematomas on her legs. There were no cutaneous stigmata of liver disease. The abdomen was slightly distended and rectal examination showed pale stools only. Examination was otherwise normal.

Investigations showed Hb 9.5 g/dl; WCC 7.6 x 10^9/l; platelets 510 x 10^9/l; MCV 90 fl; MCH 30.3 pg; MCHC 32.0 g/dl; reticulocytes 5%; iron 6 µmol/l; (normal 10-30), serum albumin 25 g/l; calcium 2.3 mmol/l; (normal 2.18-2.62); B12 320 ng/l (normal >260); folate 1.7 µg/l (normal >3); prothrombin time 120 sec (control 12); kaolin-cekaphalin clotting time (KCCT) 120 s (control 42). Clotting factors II, VII and X were all present in reduced concentration. Sigmoidoscopy was normal. Barium follow-through findings showed disease of the jejunum with a malabsorptive pattern.

The patient was treated with fresh frozen plasma and vitamin K intravenously and oral iron and folate supplements. Her prothrombin time returned to normal immediately and she showed a good reticulocyte response. She was allowed home awaiting a jejunal biopsy as an out-patient. However, soon after discharge she was admitted with tetany due to hypocalaemia, and was treated with intravenous calcium. A jejunal biopsy was performed, which showed sub-total villous atrophy, strongly in favour of coeliac disease. She was started on a gluten-free diet and was well some three months later with a normal bowel habit and haematological indices. Repeat biopsy was arranged but the patient has since left the area.

Case 2
A 21-year-old Jehovah’s Witness was admitted with a 3-day history of spontaneous bruising and bleeding per rectum. On closer questioning she admitted to occasional diarrhoea and stools which were loose and offensive. She had lost 12.7 kg in weight and had developed amenorrhoea, despite a good appetite over the previous year.

Examination showed her to be pale and thin, with multiple bruises on her arms and legs. She had a large haematoma of her right foot and a haemarthrosis of her right knee. The abdomen was distended and slightly tender.

Investigations showed her Hb to be 9.6 g/l which fell to 3.9 g/l on the second day; WCC 13.2 x 10^9/l; prothrombin time was 180 s; KCCT 180 s; plasma
fibrinogen 2.59 g/l (normal 2.0–4.0). Fibrinogen degradation products 6 μg/ml; factor II 1%, factor VII 5%, factor X 4.1%, Na+ 131 mmol/l, K+ 2.3 mmol/l, serum calcium 1.51 mmol/l, phosphate 0.72 mmol/l; alkaline phosphatase 2.7 K.A. units; albumin 23 g/l.

Vitamin K was given with reversion of the clotting screen to normal within 24 hours. The patient, however, refused blood transfusion on religious grounds and was therefore given parenteral haematinics—iron, B12 and folate together with parenteral nutrition.

A provisional diagnosis of coeliac disease was made on the basis of the history of vitamin K deficiency, hypoproteinaemia, anaemia and biochemical osteomalacia. Xylose absorption was impaired and faecal fat excretion was raised. Jejunal biopsy was performed and confirmed the presence of sub-total villous atrophy. A barium meal and follow-through examination showed no specific abnormalities.

She was allowed home on a gluten-free diet and was well some nine months later when jejunal biopsy was repeated and histology showed an improvement to partial villous atrophy. Her menstruation had returned to normal.

Discussion

Both patients who had been previously well, presented not with gastrointestinal symptoms, but dramatically in the manner described and were thought initially to have a blood dyscrasia. Review of several large series shows that although the occurrence of hypoprothrombinaemia is common, prolonged prothrombin times being found in up to 70% of untreated adult cases, (Green and Wollaeger, 1960; Bosak, Wang and Aldersberg, 1957; Barry, Baker and Read, 1974; Benson, Kowlessar and Schleisenger, 1964; Ross and Gibb, 1966), overt and severe bleeding only rarely has been reported as the initial presenting feature (Benson et al., 1964).

Although hepatic dysfunction, either due to disease or anticoagulant therapy, is the commonest cause of an acute bleeding disorder with a prolonged prothrombin time, the authors wish to emphasize that it may be the primary presenting feature in coeliac disease due to malabsorption of vitamin K.

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References


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