Case report

Brachiocephalic vein thrombosis associated with Crohn’s disease

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An association between macrovascular thrombosis and idiopathic inflammatory bowel disease has been described, although very few well-documented cases have been published. We report on a 39-year-old woman who presented with right hemifacial and upper limb edema that was shown to be due to an extensive right brachiocephalic vein thrombosis, diagnosed by magnetic resonance angiography. Laboratory findings suggested malabsorption, and a diagnosis of Crohn’s disease was established. Moreover, elevation of the plasminogen activator inhibitor system was identified. This represents the first description of a spontaneous thrombosis in a patient with Crohn’s disease involving the intrathoracic venous system and raises the possibility of impaired fibrinolysis being involved in the etiopathogenesis of this complication.

Key words: macrovascular thrombosis, Crohn’s disease, inflammatory bowel disease, thrombosis, coagulation disorders

Introduction

Inflammatory bowel disease (IBD) is a multisystemic disorder that can involve extraintestinal systems.¹ Vascular complications, although rare, have been described in association with IBD, and include both macrovascular²,³ and microvascular thrombosis,⁴ occurring predominantly during inflammatory exacerbations of intestinal disease.¹,³ Functional abnormalities in the hemostatic system have been proposed as the etiopathogenic basis for an elevated thrombogenicity in IBD,⁵,⁶ and although several of these have been investigated as potential surrogate markers for thrombotic risk, their precise role remains unknown. We report here on a patient that, to our best knowledge, represents the first description of a spontaneous macrovascular thrombosis involving the brachiocephalic vein associated with Crohn’s disease and in whom an abnormality of the fibrinolytic system was identified.

Case report

A 39-year-old Caucasian woman was admitted to the hospital because of rapidly progressive asymmetrical right hemifacial and right upper limb edema. She had a diagnosis of irritable bowel syndrome, made by her general physician after investigations that included a bowel barium enema, but she had never experienced weight loss, abdominal pain, mucus emission, or hematochezia. She was an otherwise healthy woman with two previous unremarkable gestations, at the ages of 21 and 31, and she smoked ten cigarettes a day. On admission, her temperature was 37°C, blood pressure 126/62mmHg, pulse 101, and weight, 44kg, with an index body mass of 19. There was asymmetrical edema of the right upper limb and right side of her face, with marked distension of the external right jugular vein. Cardiovascular and pulmonary examination findings were normal and there was no hepatosplenomegaly, ascites, or other abnormal abdominal findings. Hemoglobin was 10.2g/dl, white blood cell count, 5900 per mm³; platelet count, 550000 per mm³; erythrocyte sedimentation rate, 54mm/hour; and C-reactive protein, less than 0.5mg/dl. Total protein was 35g/l, with marked hypoalbuminemia (11.7g/l) and normal gammaglobulins. Prothrombin time was slightly prolonged (13.5/11s) but all other liver test results were within normal values. Calcium was 1.6mmol/l (normal, 2.1–2.6mmol/l); cholesterol, 2.49mmol/l (<5.20mmol/l); serum folates, 2.0ng/ml (4.0–18.0ng/ml); vitamin B₁₂, 180pg/ml.
(250–980 pg/ml); and serum iron, 37 µg/dl (50–170 µg/dl). Renal function tests, ionogram, urinalysis, creatine kinase, and lactic dehydrogenase findings were all normal, and antinuclear antibodies were negative. These findings suggested a malabsorption syndrome. Upper gastrointestinal endoscopy findings were normal, and a proximal jejunal biopsy showed normal villous structure and a nonspecific inflammatory infiltrate of the lamina propria. A small-bowel enema revealed a long stenosis of the terminal ileum (50 cm), nodularity, and ulceration, with cobblestone appearance and fistulous tracts in the ileocecal area (Fig. 1). Colonoscopy showed normal colonic mucosa, but the terminal ileoscopy revealed ulceration and stenosis of the ileum. Histologic findings in the colonic mucosa biopsy specimen were normal, but biopsy specimens of the terminal ileum showed epithelial atrophy and ulceration, edema, and a moderate lymphocytic and plasmocytic inflammatory infiltrate of the lamina propria. These findings led to a diagnosis of Crohn’s disease of the ileum. A chest radiograph showed small right pleural effusion. An Echo-Doppler study of the right cervical and axillary blood vessels did not reveal any abnormality, but thoracic tomographic scan showed a thrombus with total occlusion of the lumen of the right brachiocephalic vein (RBCV) with distal extension up to the superior vena cava (SVC) that was confirmed by magnetic resonance angiography (Fig. 2a), which showed a complete absence of the RBCV with a patent SVC.

Investigation of the possible mechanism of increased thrombogenicity with hemorrhheologic and coagulation studies revealed elevation of plasminogen activator inhibitor, at 15 U/ml (Table 1). These values were still elevated 1 month and 4 years after the episode (10 U/ml and 6.3 U/ml, respectively; normal range, <5 U/ml). The patient was treated with low-molecular-weight heparin 30000 U/day for 1 month, followed by warfarin at a dose adjusted by the international normalized ratio (INR), with an improvement of the upper arm and face edema. This was probably due to the development of collateral circulation and, possibly, to partial repermeation of the thrombus, because 4 years after the episode, magnetic resonance angiography revealed persistence of the thrombus (Fig. 2b).

She was also treated medically for Crohn’s disease, with mesalamine, prednisone, and metronidazole, with some improvement, but, because of subsequent small-intestinal obstruction and severe hematochezia, a resection of 60 cm of the terminal ileum was done. Pathological examination of the resected ileum re-