Case Report

Severe Clinical Course Of Ulcerative Colitis And Cerebral Venous Sinus Thrombosis Association

Deniz YERDELEN¹, Mehmet KARATAŞ¹, Özlem ALKAN²

¹Başkent Üniversitesi Tıp Fakültesi Adana Hastanesi, Nöroloji, Adana, Türkiye, ²Başkent Üniversitesi Tıp Fakültesi, Radyoloji, Adana, Türkiye

Abstract

Inflammatory bowel disease (IBD) is an idiopathic inflammatory disease of the gastrointestinal tract in which the pathogenesis is not known wholly. Thrombosis is an important risk that may be encountered in this disease and it is thought to be associated with the pathogenesis of IBD. Deep venous thrombosis and pulmonary emboli are the most common thrombotic complications in IBD patients, however, cerebral vascular involvement is rare. In these patients, although thromboembolic complications are uncommon, they are important because of their high mortality. In this report, the clinical picture with a fatal course of a patient, newly diagnosed with ulcerative colitis, who developed a cerebral venous sinus thrombosis is discussed.

Keywords: ulcerative colitis, cerebral venous sinus thrombosis

INTRODUCTION

Ulcerative colitis (UC) is an idiopathic inflammatory bowel disease. The exact pathogenesis of UC is unknown. However, genetic susceptibility and environmental influences have been proposed in the pathophysiology of UC. Thrombosis in patients with UC is a well-known risk, and this pro-thrombotic state in UC contributes to the pathogenesis of this condition. Therefore, various extra-intestinal manifestation such as thromboembolic disease may develop. Most thromboembolic events occur in the lower extremities and pelvis, whereas the incidence of central nervous system involvement is rare and varied (6,11). In UC, thromboembolic
complications are uncommon, but are important because of their high mortality (12,13). Here, we presented a 20-years-old woman with severe neurological signs and symptoms associated with superior sagittal sinus thrombosis, and venous infarct, newly diagnosed with UC to emphasize that this complication of UC may be devastating in spite of treatment, especially if the neurological clinical picture is severe.

CASE PRESENTATION

A 20-year-old woman was admitted to our clinic with sudden onset impaired consciousness and left-sided weakness. One week prior to presentation at our clinic, while being investigated for hemorrhagic diarrhea continuing for 4 to 5 months, she had been diagnosed as having UC. A physical examination revealed the following: body temperature, 39°C; blood pressure, 110/70 mm Hg; and pulse, 127 beats/minute. A neurologic examination demonstrated lethargy and left-sided hemiplegia. The results of standard laboratory analyses revealed mild anemia (hemoglobin, 11 g/dL and hematocrit, 33%) and mild leucocytosis (white blood count, 13,400 K/mm³). The erythrocyte sedimentation rate was mildly elevated (25 mm/h) and the C-reactive protein level was prominently elevated (96 mg/dL). The thromocyte level, prothrombin time, activated partial thromboplastin time, coagulation factors V and VIII, fibrinogen, protein C, and anticardiolipin antibody IgM and IgG levels were normal; the protein S level was low (13; normal range, 61-91); and the results of antinuclear antibody and anti-dsDNA tests were negative. Antithrombin 3 was mildly elevated (136, normal range: 75-125). The results of liver, renal, and thyroid function tests and urine analyses were normal. Cerebral magnetic resonance imaging showed a superior sagittal sinus thrombosis and a venous infarct with a hemorrhagic component, which caused a right frontal subfalcine herniation (Figures 1 A and B). The patient had secondary generalized convulsions on the second and third days of hospitalization. An electroencephalography demonstrated right-sided, paroxysmal, lateralized, epileptiform discharges. Low molecular weight heparin, acetyl salicylic acid, and (after the convulsions) phenytoin were started, and the sulfasalazine that she had been taking was continued. On the 14th day after admission, her temperature and pulse returned to normal values, she became conscious, and left-sided hemiplegia continued. However, the number of defecations increased, and so prednisolone (40 mg/day) was added to the treatment by a gastroenterologist. On the 20th day of hospitalization, respiratory deficiency and tachycardia developed. Blood gas findings showed hypoxia and hypocapnia. A thoracic computed tomography scan performed to exclude pulmonary emboli revealed pulmonary edema. Her central venous pressure was 30 cm. Echocardiography did not show any pathological findings. She was diagnosed as having pulmonary edema and in spite of treatment, she died on the same day of this condition.

DISCUSSION

Thromboembolism is a known but rare complication of inflammatory bowel disease (IBD) occurring in 1.3% to 6.4% of all such patients and in up to 39% of autopsies (7,11). The pathogenesis of thromboembolism in UC remains unclear. Present data indicate that thrombosis in IBD multifactorial (13). Suggestions for the high prevalence of thromboembolism in IBD patients include prothrombotic factors (anti-cardiolipin antibodies, hyperhomocysteinemia, and changes in the lipid spectrum), IBD-induced alterations in plasma and mucosal hemostasis (increased endothelial activation and tissue factor
expression, and impaired inhibition of coagulation in IBD), and changes secondary to inflammation. The balance between procoagulant and profibrinolytic factors in IBD patients seems to favor plasma hypercoagulability. The coagulation abnormalities associated with active IBD include elevated levels of factors V and VIII and fibrinogen, decreased levels of antithrombin III, as well as quantitative and qualitative platelet disorders. Hyperhomocysteinemia has been investigated, and although an elevated total homocysteine level is common in IBD patients, data demonstrate that this factor does not contribute to the development of thrombotic complications. In a reported study, the higher prevalence of thromboses in IBD patients was not found to have a genetic linkage. Hereditary conditions predisposing to thrombosis such as factor V mutation, prothrombin gene mutation, MTHFR mutation, proteins C and S, antithrombin III deficiencies, and IBD were not found to be correlated.

Deep venous thromboses and pulmonary emboli are the most common thrombotic complications in IBD patients; however, cerebral vascular involvement is rare. Cerebral vascular involvement, although uncommon, may occur as cerebral venous and dural sinus thromboses. The superior sagittal sinus and lateral sinuses are commonly involved. Cerebral (dural) venous sinus thrombosis is an uncommon condition, often affecting young to middle-aged patients and more frequently women. Although known for more than 100 years, it is only because of greater awareness among physicians and...
neurologists and by improved noninvasive imaging techniques that has cerebral venous sinus thromboses have been diagnosed prior to death in recent years (5).

The clinical presentation of cerebral venous sinus thrombosis can be extremely varied and include headache (95%), focal seizures with or without secondary generalization (47%), paresis (uni- or bilateral) (43%), papilledema (41%), impairment of consciousness (54%), isolated intracranial hypertension (20%), and type picture (headache, visual disturbance and papilledema) (1).

Thunderclap headache mimicking subarachnoid hemorrhage has also been reported (2).

Severe neurologic clinical pictures (eg, hemiparesis) due to cerebral venous sinus thromboses associated with UC with more severe courses have been reported (13). However, mild neurologic involvement like headache, papilledema, sixth nerve paresis, mild monoparesis, and mild seizures may respond better to treatment and have a better clinical course (3,7,10,12).

Our patient presented with severe neurologic signs and symptoms, and she had a history of newly diagnosed UC. Treatment for both conditions began, and during minimal improvement of the neurologic signs, the symptoms of UC intensified. Our patient’s severe course may be explained as follows: The inflammation-induced changes resulted in coagulation activation, which induced proinflammatory effects so that a looping chain of reactions similar to that reported in the literature became manifest ultimately leading to the death of this patient (14).

By presenting this case, we wish to emphasize that during the initial period of UC, patients may present with neurologic symptoms and signs, and cerebral (dural) venous sinus thromboses should be considered as a cause of this clinical picture. In instances of severe neurologic findings, the clinical outcome may be fatal.

Correspondence to:
Özlem Alkan
E-mail: alkano@yahoo.com

Received by: 29 July 2007
Revised by: 07 September 2007
Accepted: 29 October 2007

REFERENCES