Pulseless Hematochezia: Takayasu’s Arteritis Associated with Ulcerative Colitis

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Abstract

A 36-year-old woman with ulcerative colitis presented with fever, chest and back pain, and fatigue sensation of the arm. Her upper limb pulses were absent. Angiography showed multiple aneurysms of the aorta and its branches, consistent with Takayasu’s arteritis. She showed HLA-B35 but no B52, which is the typical haplotype among the coexistence cases of both diseases. Prednisolone was effective. The possible pathogenic association of the disorders is discussed.

Key words: Takayasu’s arteritis, ulcerative colitis, HLA

Introduction

Ulcerative colitis (UC) is often complicated with other autoimmune diseases. Takayasu’s arteritis (TA), also known as “pulseless disease”, is a presumed autoimmune vasculitis mainly involving the aorta and its main branches (1). The coexistence of UC and TA has rarely been reported. We report an unusual case of a patient who had initially suffered from UC and subsequently developed TA.

Case Report

A 36-year-old Japanese woman, having a history of UC and pyoderma gangrenosum for nine years, was referred on September 17, 1985 for exacerbation of hematochezia as well as fever, chest and back pain, and fatigue sensation of her left arm. On admission, both upper limb pulses were absent. Blood pressure was 150/60 mmHg at the left popliteal artery. Levine grade II/VI systolic and diastolic murmurs were audible along the left sternal border. Her abdomen was soft with hyperactive bowel sounds and moderate tenderness to palpation in the left lower quadrant, but no bruits. Laboratory parameters showed hemoglobin 4.5 g/dl, leucocytes 13,500/mm³, and erythrocyte sedimentation rate 160 mm/h. HLA typing was A2, B35, and Cw3. Barium enema examination showed multiple pseudopolyps extending from the rectum to ascending colon. The absence of hastra resulted in a tubular, rigid narrowed appearance (Fig. 1). Sigmoidoscopy revealed diffuse hemorrhagic friable mucosa with loss of vascular pattern and pseudopolyps, consistent with UC. Catheter angiography showed aneurysm formation of the ascending aorta, aortic arch, thoracic descending aorta, and bilateral common carotid arteries (Fig. 2). It also demonstrated left subclavian aneurysm, aortic regurgitation (Sellers’ grade I) and intact coronary arteries. TA was diagnosed on the basis of the valid criteria. Considering multiple aneurysm formation of the major arteries by active inflammation, cardiovascular surgery was not indicated at that time. The antihypertensive treatment with nifedipine (40 mg/day), labetalol (150 mg/day), and isosorbide dinitrate (40 mg/day) was initiated. Her bowel and cardiovascular condition were resolved gradually with prednisolone (60 mg/day). As an adjunct to prednisolone, metronidazole (250 mg/day) was effective for UC. Follow-up radiographic studies revealed no change of her aneurysms, however, her radial pulses gradually recovered in 3 years. Her condition remained uneventful with stable blood pressure and cardiac function for 13 years of follow-up until a sudden lethal aneurysmal rupture on December 14, 1998.

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Discussion

TA produces marked fibrosis and thickening of the arterial wall resulting in occlusion of the major arteries, which presents with various ischemic symptoms of upper extremities and cervical lesions (1). In general, thoracic aneurysms of greater than 6 cm in diameter are prone to rupture, thus elective surgical procedures including vascular grafts should be indicated. However, especially for those lesions involved in TA, treatment must be individualized with higher risks of anastomotic complications in affected major arteries (2). The present patient chose to avoid surgery with informed operative advantages and risks of extensive aneurysmal involvement.

The pathogenic association of TA with UC is not clear, however, a common genetic basis has been speculated because of the high frequency of specific HLA-B52 and DR2 for the two disorders among Japanese patients (3). It is a novel finding that a Japanese patient with both diseases carries no B5 or B52, as in the present case. The coexistence of UC and TA in this case may be coincidental, or due to cross-reactivity between autoantigens in the arterial wall and colonic mucosa. Recent studies support a role for bacteria in the pathogenesis of UC (4), whereas researchers of the vascular pathology have put forward the hypothesis that microbes or their structural components with high sequence homology to humans, may initiate and accelerate chronic infection, induce autoimmunity against vascular cells, and lead to an atherosclerotic process (5). One might think that certain microorganisms provoke chronic autoimmune diseases at both the arterial wall and colonic mucosa of genetically susceptible individuals. In conclusion, TA may accompany UC as an extraintestinal manifestation.

References