Ulcerative Colitis Associated with Takayasu’s Disease

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A 25-year-old Japanese woman had both ulcerative colitis and Takayasu’s disease and was positive for HLA-A24, BW52, and DR2. She was found to have thickening of the wall of the carotid artery on contrast-enhanced computerized tomography of the neck and chest. Prednisolone, beraprost, and sulfapyridine achieved rapid remission of both diseases.

Key words: ulcerative colitis, Takayasu’s disease, HLA-BW52, HLA-A24, contrast-enhanced computed tomography (CE-CT)

Introduction

Ulcerative colitis (UC) complicated by Takayasu’s disease has been considered to be extremely rare. We however, recently encountered three patients with these combined diseases. The first one, an autopsy case, was previously reported, with a review in the available literature (1). The present report describes the second patient with UC and Takayasu’s disease. The third is submitted for publication. We discuss the usefulness of measuring MHC class I and II type for diagnosis of the combination of UC and Takayasu’s disease, taking into account also contrast-enhanced computed tomography (CE-CT) of the neck and chest to monitor the intensity of the arteritis.

Case Report

A 25-year-old Japanese female dentist complained of fever and anterior cervical pain in April 1991. She was admitted to a local hospital with an initial possibility of subacute thyroiditis. Later, Takayasu’s disease was suspected because of a weakness in her left pulse tension and a bulging of the left carotid artery with bruit. Thereafter, she was transferred to the Department of Neurology and Neurosurgery, Fukushima Medical College for further examination.

The aortography done in May 1991 (Fig. 1) showed that the proximal region of the left subclavian artery was interrupted together with the distal part which was supplied by the bypass from the left vertebral artery. Stenosis accompanied by some dilatation was observed in the left common carotid artery and also in the aorta just above the celiac artery. Based on these findings, Takayasu’s disease was diagnosed, and she was treated with prednisolone (PSL) in an initial dose of 50 mg/every other day (E.O.D.), which was tapered to 15 mg/E.O.D., and ticlopidine hydrochloride at a dosage of 75 mg/day.

In October, 1992, she complained of an attack of photophobia and pain in both eyes, which was ophthalmologically diagnosed as acute iritis. A few days later, she expelled bloody mucus two to five times a day, accompanied by an intermittent pain in the left lower abdomen.

Proctosigmoidoscopic examination revealed reddish, edematous and fine granular mucosa with muco-purulent exudate and oozing blood in the rectum (Fig. 2), which was diffusely and continuously extended to the sigmoid colon. Biopsy specimen of the rectal mucosa showed a chronic nonspecific inflammation consisting of regenerated glands, goblet cell depletion and lymphoid cell infiltration, but there were no findings of crypt abscess or ghost appearance.

Barium enema examination showed thumbprint and “collar-button” configurations in the sigmoid colon as well as fine spicula and coarse flecks of barium in the rectum (Fig. 3). No abnormal findings were observed in the descending, transverse and ascending colon.

Inquiry into her previous health record revealed attacks of diarrhea with bloody mucus and lower bowel discomfort, which continued over a month at the age of 24 and 25. Her bowel symptoms were thought possibly due to “ischemic colitis” associated with Takayasu’s disease. However, angiography of the superior and inferior mesenteric artery showed nothing abnormal.
Fig. 1. Aortogram in May 1991 showed stenosis and dilatation of thoracic to abdominal aorta just above the celiac artery, stenosis and dilatation of the common carotid and an interruption at the proximal region of the left subclavian artery.

Fig. 2. Proctoscopic photograph in Oct. 1992 showed reddish, edematous and fine granular mucosa with mucopurulent exudate and oozing blood in the rectum.

Fig. 3. Roentgenogram in Oct. 1992 demonstrated thumbprint and "collar-button" configurations in the sigmoid colon, and fine spicula and coarse flecks of barium in the rectum.

Based on these endoscopic and roentgenographic findings, an additional diagnosis of ulcerative colitis was arrived at and she was transferred to our department.

Physical findings on Oct. 7, 1992 were the following; Photophobia and pain of her eyes improved without any sequela. Blood pressure of the brachial artery was 110/mmHg in the left, as no diastolic pressure appeared, and 122/78 mmHg in the right. Pulse tension was weak in the left upper limb and normal in the other limbs. Her left arm felt colder than the right. A bruit was audible on the left neck and around the clavicle, but not in the heart region nor abdomen.

Laboratory data were as follows: erythrocyte sedimentation rate 42 mm/hr, CRP 2.9 mg/dl, white blood cell count 13,000/mm³ (72% neutrophils, 24% lymphocytes and 4% monocytes), red blood cell count 427×10⁶/mm³, platelet count 30.8×10⁴/mm³, prothrombin time and active partial thromboplastin time were within normal limits; blood chemistry showed normal hepatic and renal function; antinuclear antibody, rheumatoid factor and lupus anticoagulant were negative; HLA-typing was positive for A24(9), A33(19), BW52(5), BW44(12), DR2, and DR6.

Elevated levels of ESR, CRP and leukocyte counts were considered, in part, as a result of a flareup of UC, while the activity of Takayasu’s disease could not be evaluated. Therefore, she received a low-residue diet and was treated with a large-dosage of PSL (50 mg/day) and beraprost sodium (prostaglandin I₂, 60 μg/day). Salazosulfapyridine (SASP) was not initially administered, because of our previous experience with a worsening of the symptoms of Takayasu’s disease observed in a 14-year-old female patient with both UC and Takayasu’s disease (submitted for publication). CE-CT scan of the neck and chest taken on Oct. 15 showed a thickening of the wall of the left carotid artery with a luminal narrowing (Fig. 4). In addition, ¹³¹I-macro-aggregated albumin scintigram revealed
Fig. 4. Contrast-enhanced CT of the chest in Oct. 1992 showed a thickening of the wall of the left carotid artery (arrowhead) with a narrowing of the lumen (arrow).

a decrease of the blood-flow in both lungs.

Diarrhea with bloody mucus and bowel pain improved within 3 weeks after the treatment with PSL and beraprost. Then, SASP was carefully administrated just before tapering PSL from 30 mg/day to 25 mg/day. Her bowel symptoms became quiet and laboratory findings for inflammation improved rapidly without aggravation. She was discharged on Nov. 20, 1992.

Discussion

The incidence and prevalence rates of UC in Japan are reported to be 0.36/100,000 in 1980's (2) and 7.85/100,000 in 1985 (3), respectively, that is to say nearly 1/10 to 1/20 of the rates in Western countries (4–6). On the other hand, Takayasu's disease is more common in Japan than in the Western countries. Nasu disclosed pathological findings of Takayasu's disease in 100 cases out of 293,315 general autopsies (0.04%) in Japan (7). However, figures for overt Takayasu's disease are not high even in Japan, since most early cases of Takayasu's disease lack subjective symptoms.

An overlap of UC with Takayasu's disease has been considered to be a fairly rare condition and only thirteen Japanese cases have been reported from 1964 to 1988 (1). During the past two years, however, we have encountered three patients with both UC and Takayasu's disease in our department. All three patients were Japanese females and showed HLA-A24 and BW52, and two of them additionally showed DR2 phenotype in (submitted for publication). The frequent incidence of HLA-A24, BW52 and DR2 as observed in these three patients with both UC and Takayasu's disease has also been encountered in other Japanese cases (1). The male/female ratio, including our three cases, was 2/13, and all of seven cases, in which HLA was typed, showed either HLA-BW52 or B5 (1). The relative risk of HLA-BW52 was 5.26 for UC (8) and 5.5 for Takayasu's disease (9), respectively. Therefore, a few patients who have UC and HLA-BW52 would be suspected of latent Takayasu's disease.

CE-CT scan might be recommended for an early diagnosis of Takayasu's disease rather than angiography or magnetic resonance imaging (MRI), since CT is more readily available and less stressful than the other exams. It also gives a clear image of a narrowed lumen and a thickened wall of the affected artery in the contrast-enhanced condition, as was observed in this patient.

Finally, the following should be emphasized. A combination of ulcerative colitis and Takayasu's disease may be a type of an overlap syndrome associated with genetic factors. A contrast-enhanced computed tomography (CE-CT scan) of the neck and chest offers an easy and effective method for the diagnosis of Takayasu's disease.

References