Takayasu’s Disease in a Patient with Ulcerative Colitis

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A 35-year-old Korean man with a 10-year history of ulcerative colitis (UC) presented with pain and swelling of the right neck. The patient was diagnosed with Takayasu’s arteritis (TA) and had human leukocyte antigen (HLA) B-52, which is frequently found in patients having both UC and Takayasu’s disease concurrently on HLA analysis. This case is the first report of a patient with both TA and UC in Korea, to the best of our knowledge. ([Korean Circ J 2013;43:135-138])

KEY WORDS: Takayasu arteritis; Colitis, ulcerative; Human leukocyte antigen-B52 antigen.

Introduction

Takayasu’s arteritis (TA) is a chronic vasculitis of uncertain etiology affecting large and medium-sized arteries, primarily the aorta and its branches as well as the coronary and pulmonary arteries. This disease is not endemic to a particular area, but is developed mostly in young women of Asia. It has been reported that about 150 new cases develop annually in Japan, but in the United States and Europe, 1 to 3 cases per 100,000 persons have occurred per year. Ulcerative colitis (UC) is an idiopathic chronic inflammatory disease limited to the mucosa and submucosa of the colon. UC almost always involves the rectum and may extend to other parts of the colon. The incidence of UC has been reported to be high in Northern Europe, the United Kingdom, and North America. The prevalence of UC has been estimated to range from 37 to 246 cases per 100,000 persons in North America. The occurrence of TA in patients with UC has been reported rarely with only about 50 cases in the world and more than half of these cases have been identified in Japan. A case of this type has not been reported in Korea until now, to our knowledge. Herein, we report a middle-aged Korean man with both TA and UC.

Case

A 35-year-old man visited our outpatient clinic in June 2010, presenting with pain and swelling of the right neck for 6 months. He was diagnosed with UC at his age of 25 years and had taken 2250 mg of mesalazine daily.

His chest X-ray was normal, but thyroid sonogram showed severe thickening of both carotid intima and media, and ulceration at the mid-portion of the right carotid artery (Fig. 1). Carotid CT angiography showed concentric wall thickening of both carotid arteries (Fig. 2). Echocardiogram showed no specific findings.

Laboratory data were as follows: white blood cell count, 9550/mm³ with 67.9% neutrophils; hemoglobin, 8.73 g/dL; mean corpuscular volume, 57.2 femtolitre; mean corpuscular hemoglobin concentration 29.7%; erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were high at 69 mm/h and 107.5 mg/L, respectively.

The occurrence of TA in patients with UC has been reported rarely with only about 50 cases in the world and more than half of these cases have been identified in Japan. A case of this type has not been reported in Korea until now, to our knowledge. Herein, we report a middle-aged Korean man with both TA and UC.
ported a claudication of the right upper extremity during use. Bruits were audible over the right carotid and left subclavian artery area. Coronary angiography showed no remarkable findings. However, arteriographic narrowing of the aortic branches was diffuse, especially in the right and left external carotid artery (Fig. 3). Chest CT angiography showed concentric wall thickening of the ascending thoracic aorta, aortic arch, proximal descending thoracic aorta, as well as the innominate artery, left common carotid, and both subclavian arteries (Fig. 4). He was diagnosed as concurrent TA and UC based on these findings and his history.

The patient received prednisolone (30 mg/day) and ferrous sulfate (256 mg twice a day) with mesalazine. Since then, several follow-ups were carried out during the following months and the patient’s symptom included right neck swelling and improvement of pain. CRP and ESR levels declined to 24 mm/h and 14.2 mg/L, respectively. Hemoglobin levels were normalized (14.6 g/dL).

During this follow-up period, immunological studies were conducted for evaluation of the patient’s autoimmunity. Laboratory data showed: auto antibodies including antinuclear, anti-neutrophil cytoplasmic, anti-DNA and antiphospholipid antibodies, which are associated with other vascular diseases, but not TA were negative.

Human leukocyte antigen (HLA) studies showed B51 and B52, which are usually seen in cases with coexisting UC and TA. In addition, the patients underwent a colonoscopy. There was moderate pancolitis and a polyp (7 mm) in the ascending colon (Fig. 5). A polypectomy was performed and a biopsy showed no dysplasia. With the pa-

Fig. 1. Thyroid sonogram of both carotid arteries shows severe thickening of the right (A) and left (B) carotid intima and media walls (arrows), in addition to right carotid artery ulceration at the midportion (A).

Fig. 2. Carotid CT angiography shows concentric wall thickening of the right (A) and left (B) common carotid artery (arrows).
tient's improved state, prednisolone was tapered down and the pa-
tient has been monitored through the outpatient clinic.

Discussion

Takayasu's arteritis is a chronic vasculitis of unknown etiology, and usually seen in young Asian women. TA, which primarily involves the aorta and its major branches, often occurs initially in the left middle or in the proximal subclavian artery. During the development of disease, both of the common carotid and vertebral arteries, the brachiocephalic artery, the right middle or proximal subclavian artery, and the aorta may be involved. In about 50% of patients, the abdominal aorta and pulmonary arteries are affected.

With regard to diagnosis, criteria for the classification of TA have been suggested in 1990 by the American College of Rheumatology (ACR) as: 1) onset at age less than or equal to 40 years. 2) claudication of the extremity. 3) decreased pulsation of one or both brachial arteries. 4) greater than a 10 mm Hg difference in systolic blood pressure between arms. 5) bruit over the subclavian arteries or the aorta. 6) arteriographic evidence of narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities. The presence of 3 or more of these 6 criteria demonstrated a sensitivity of 90.5% and a specificity of 97.8%. In this case, the patient had an occasional right upper extremity claudication, but did not have abnormal blood pressure of the right arm. However, for this patient, disease onset occurred at 35 years and he had bruits of the left subclavian artery. In addition, the patient's arteriogram was abnormal. Therefore, our patient met the ACR criteria. Additionally, the patient had tenderness of the carotid artery and had an elevated ESR and anemia, which are important evidence of TA.

Ulcerative colitis, a form of inflammatory bowel disease (IBD), is characterized by relapsing inflammation affecting mostly the mucosal layer and sometimes the submucosal layer of bowel. In gen-

Fig. 3. Arteriogram of carotid arteries shows narrowing of the left external carotid artery (arrow).

Fig. 4. Chest CT angiography shows concentric wall thickening of the innominate, left common carotid and both subclavian arteries.

Fig. 5. Colonoscopy shows a polyp at ascending colon (A), and erythematous, edematous mucosa with granularity, friability at the descending colon (B).
eral, UC occurs in North America and Europe, but the incidence has increased in Asia and in other countries, including Japan, Korea, Singapore, and Latin America, inferring that industrialization may affects the occurrence of UC. UC and Crohn's disease (CD), the 2 major types of IBD are sometimes related with other autoimmune diseases and extraintestinal symptoms, including vascular manifestations.

As seen above, the prevalent geographical areas of UC and TA are different, but an overlap of these 2 diseases has been reported in Sri Lanka, India, Pakistan, Turkey, and especially in Japan. In Korea, a case of CD associated with TA was reported, but this is the first case of coexistent UC and TA.

With respect to an autoimmunologic association on the coexistence of UC and TA, HLA B52 and DR2 have been found in many Japanese cases in which the patient suffered from concurrent UC and TA. Indeed, our patient was positive for HLA B52, which is the common haplotype seen in cases with both diseases. In recent years, successful treatment of a patient with refractory active TA complicated by UC using anti-interleukin-6 receptor antibody was reported. In patients with these 2 diseases, there is a tendency that UC is diagnosed earlier than TA. The cause of this tendency is unknown, but there are suggestions that bacterial or viral invasion through the intestinal mucosa may be associated with the pathogenesis of these cases.

Like this patient, there are a few cases presenting both diseases even though the possibility is very low from an epidemiological point of view. In this sense, it is possible that they are immunologically related. In other words, considering that anti-colon antibodies and anti-aortic antibodies have been demonstrated in UC and TA, we cannot exclude a possibility that some immunological defect might exist simultaneously for both diseases, or we can hypothesize that certain microorganisms trigger chronic autoimmune disease associated with the arterial wall and colonic mucosa of immunologically susceptible individuals. Among these conjectures, this case could be another instance where an immunological and genetic link might exist between these two diseases, and it could be important for a patient having UC in Korea that careful assessment of extraintestinal manifestations should be done with a consideration of autoimmune and vascular diseases including TA.

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