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LETTER

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Takayasu arteritis associated with systemic sclerosis

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Takayasu arteritis (TA), or pulselessness disease, is a necrotizing and obliterative giant cell arteritis whose etiology is unknown; it involves the aorta and its main branches, and coronary and pulmonary arteries. It is diagnosed by clinical findings and angiographic investigation.[1] Systemic sclerosis (SSc) is a connective tissue disease of unknown etiology. Its vasculopathy is manifested by abnormal collection of collagen and extracellular matrix in skin and internal organs, and is caused by immunologic mechanisms.[1] Causes of TA associated with SSc are rarely found.[2] We report the case of a patient with TA who developed SSc 1 year later. This is the third case of Takayasu arteritis associated with systemic sclerosis reported in the literature.

A 48-year-old female patient was admitted to our clinic with complaints of malaise and cyanosis on the fingertips, especially in cold environments. One year previously a physical examination for intermittent claudication had shown that she had no pulse on the distal arms. Doppler ultrosonographic investigation showed 85% and 75% narrowing of the right and left subclavian arteries, respectively. The narrowing showed that the left and right subclavian arteries were obliterated on the distal side of the vertebral artery bifurcation region (Fig. 1). Angiographic investigation of the lower extremities revealed that visualization was not possible for the right dorsal pedal artery, the 1/3 distal side of left tibial anterior artery, and the left dorsal pedal artery. The patient was diagnosed as having TA. She had a 3-year history of Raynaud phenomenon in her medical record. Physical examination showed that her blood pressure was 150/70 mmHg on right thigh, pulse rate rhythm 74/min, and upper extremity pulses could not be palpated. There was no murmur on the vessels of the neck. There were dermal indurations on the proximal metacarpopharyngeal joints and fingers of both hands. There were no abnormalities on the other systemic organs.

Laboratory investigations were as follows: leukocyte 10500/μl, neutrophil 8100/μl, lymphocyte 1400/μl, erythrocyte 4000×10^3/μl, hemoglobin 12 g/dl, hematocrit 35%, mean erythrocyte volume 91 fl, thrombocyte 223000/μl, erythrocyte sedimentation rate 14 mm/h, and CRP 7.2 mg/l; serum biochemical examination showed fasting glucose intolerance. Human leukocyte antigen (HLA) tissue typing by Terasaki microphototoxicity method showed HLA-A1, 11; HLA-B*41, 52; HLA-DRB1*08, 10. Her thyroidal hormone profile was normal and HbA1c level 6%. Serologic investigation revealed that she had positive nuclear type antinuclear antibody and positive anticentromer antibody in 1/320 dilution, and positive anticardiolipin IgM 14.4 MPLU/ml (normal, <40), anticardiolipin IgG 10.4 MPLU/ml (normal, <11) antibodies. Her chest radiography, echocardiography, and respiratory function tests were normal.

The patient was diagnosed as having systemic sclerosis by the findings of sclerodactyly, the presence of Raynaud phenomenon for 3 years, and positive anticentromer antibody. Diagnostic clinical evidence on SSc included the finding of Raynaud phenomenon and the presence of a short period between Raynaud phenomenon and SSc, and symmetrical skin involvement in both forearms, face, and trunk, as well as narrowing of the mouth orifice. The supporting serological evidence included nuclear type ANA staining and positive anticentromer antibody. She was given calcium channel blocker in addition to Takayasu arteritis therapy.

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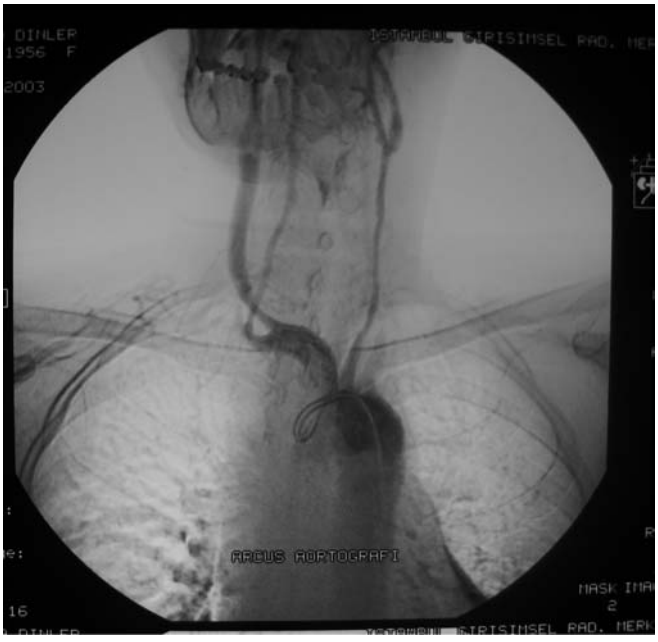

Fig. 1. Narrowing showed that the left and right subclavian arteries were obliterated on the distal side of the vertebral artery bifurcation region

Takayasu arteritis has been found to be associated with HLA-B3902, B5201, DRB1*1502, DPB1*0901 by HLA-B gen and class 2 gen (DRB1, DQA1, DQB1, DPB1) typing in Japanese people, HLA-B5 in Indian people, and HLA-B52 and HLA-B15 in Mexican Mestizo people.[1–3] Progressive systemic sclerosis is associated with HLA antigens B8, DR3 and DR52.[4] In addition, it is also associated with HLA antigens A9 and its subgroup Aw24.[5] Systemic sclerosis is found to be strongly associated with HLA-DRB1*1104 allele in the Greek community.[6]

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