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Takayasu's arteritis in a case diagnosed as Crohn's disease

Crohn hastalığı tanılı bir olguda Takayasu arteriti

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Takayasu's arteritis is a chronic granulomatous vasculitis of unknown etiology that primarily affects the great arteries; the aorta and its branches, and pulmonary and coronary arteries are the most commonly affected. Takayasu's arteritis and inflammatory bowel disease in the same individual has been occasionally reported in the literature. We report a case with Takayasu's arteritis and Crohn's disease.

Key words: Inflammatory bowel disease, Takayasu's arteritis

Takayasu arteriti (TA) primer olarak büyük arter tutulumu ile seyreden etyolojisi bilinmeyen, kronik granülamatöz bir vaskülittir. Aorta ve dalları, koroner ve pulmoner arterler en sık tutulan damarlardır. Takayasu arteriti ve inflamatuvar barsak hastalığının (İBH) aynı bireyde görülmesi literatürde nadir olarak rapor edilmiştir. Bu yazıda Crohn hastalığı ve Takayasu arteriti tanılı bir olgu sunulmuştur.

Anahtar kelimeler: İnflamatuvar barsak hastalığı, Takayasu arteriti

INTRODUCTION

Crohn's disease (CD) is a chronic granulomatous inflammatory disease that can affect any region in the gastrointestinal system from mouth to anus. With respect to the large series, the incidence and prevalence of CD are reported as 23/100,000 and 199/100,000 (2). The incidence of Takayasu's arteritis (TA) is estimated as 1-3/1,000,000. The probability of these two diseases developing in the same individual is $1/10^{10}$. This relationship, which is unexpected theoretically, was reported in children, adults and pregnants in the literature (4-14). We report our case, who was followed up with CD and was subsequently diagnosed as TA. We also discuss the probable pathophysiologic mechanism between the two diseases.

CASE REPORT

A 25-year-old female patient admitted to the infectious diseases clinic of our hospital five months previously with the complaints of widespread 10-15 mm erythematous rash over the whole body, fever, cough and diarrhea. She diagnosed as CD with colonoscopy and biopsy. She was discharged from the hospital with mesalazine 4 g/day treatment. While she was being followed in the outpatient clinic with azathioprine 2 mg/kg and mesalazine treatment, she had headache, back, knee and ankle pain and stomachache. Physical examination revealed arterial tension of 140/80 in the right arm and 110/60 in the left arm. The radial artery pulse was 90/minute on the right side and lower on the left. Her fever was 38.5°C. She also had restriction in back movements and hip abduction as well as pain with hip abduction. Laboratory investigations revealed hemoglobin (Hb): 10.6 g/dl, hematocrit (Hct): 33.6%, mean corpuscular volume (MCV): 90.7 fl, erythrocyte sedimentation rate (ESR): 90 mm/hour, C-reactive protein (CRP): 12.1 (0.00-0.800) mg/dl, albumin: 3.3 g/dl, globulin: 4.4 g/dl, iron (Fe): 15 mg/dl, and total iron-binding capacity (TIBC): 171 II g/dl. In clinical follow up, we suspected tuberculosis because of persisting fever, anemia and high sedimentation rate and CRP. Hence, colonoscopy was repeated, revealing deep

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ulcers with cobble-stone appearance and patchy distribution throughout whole segments of the colon correlated with CD. In the biopsy findings, cryptitis, crypt distortion, focal goblet cell loss, and deep linear ulcers were found and these findings were in accordance with CD. No tuberculosis bacilli could be determined in the endoscopic biopsy specimen with polymerase chain reaction (PCR) technique or acid bacilli on histological examination. The patient was consulted with Rheumatology and pelvis radiography demonstrated bilateral subchondral sclerosis and irregularity in surfaces of sacroiliac joints (Figure 1). HLA B27 Ag was determined and we therefore considered seronegative spondyloarthropathy. In ophthalmology consultation, uveitis was determined in the left eye. Power Doppler examination of upper and lower extremities was done because of arterial tension difference between right and left arms. We found monophasic, low amplitude flow in all arteries (from axillary artery to radial and ulnar arteries) of left upper extremity (Figures 2, 3, 4, 5). Bilateral carotid and vertebral artery power Doppler analysis demonstrated intimal thickening in bilateral common carotid artery (CCA), and internal (ICA) and external carotid artery (ECA). In echocardiography, minimal aortic and mitral valve insufficiency was found. We performed aortography and angiography which demonstrated a plaque in the proximal part of the ECA, causing 50% obstruction (Figure 6). With the criteria of young age, female patient, no brachial pulse, and pulse pressure difference of more than 10 mmHg between the two upper extremities and based on angiography findings, we considered TA according to the criteria of the 1990 American College of Rheumatology (ACR) classification. We started steroid 20 mg/day and methotrexate 15 mg/week therapy and discharged the patient. In the first month follow-up, Hb was 12.8 g/dl, Hct 40%, sedimentation rate 29 mm/hour, and temperature 36°C.

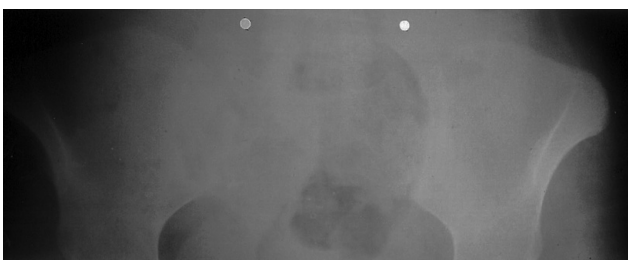
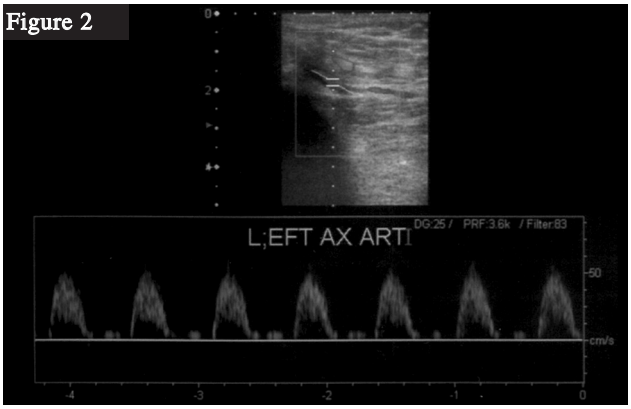
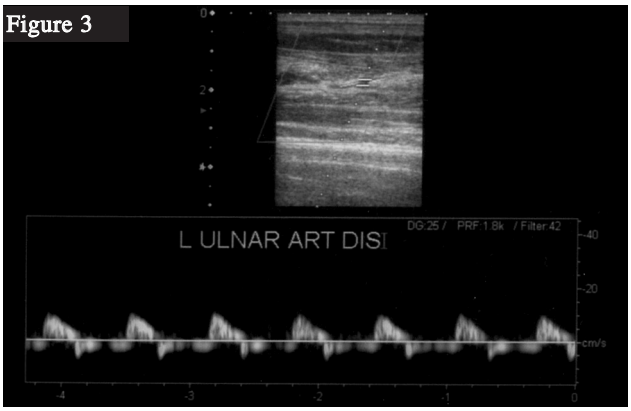
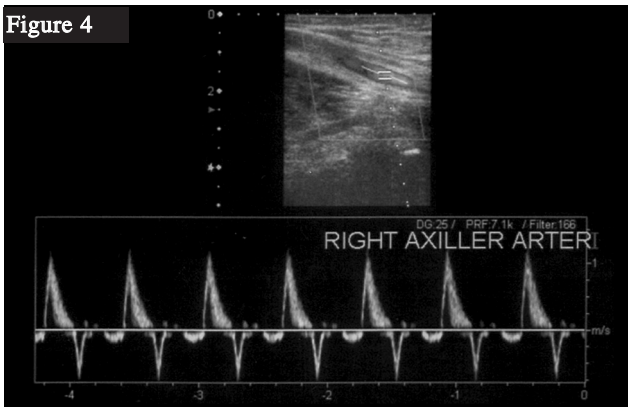


Figure 1. Bilateral subchondral sclerosis and irregularity in surfaces of sacroiliac joints in the pelvis radiography







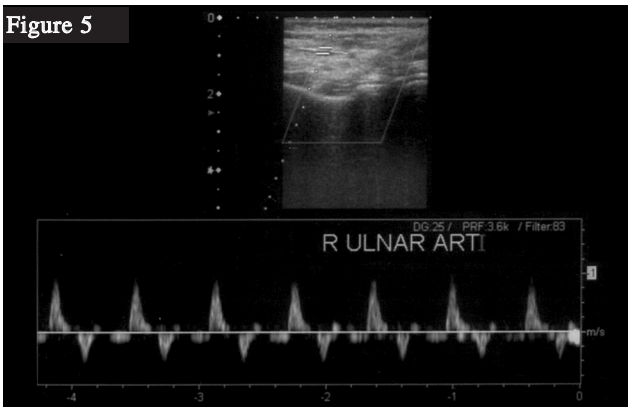


Figure 2, 3, 4, 5. Monophasic, low amplitude flow in all arteries (from axillary artery to radial and ulnar arteries) of left upper extremity and triphasic, high amplitude flow in all arteries (from axillary artery to radial and ulnar arteries) of right upper extremity in the power Doppler examination

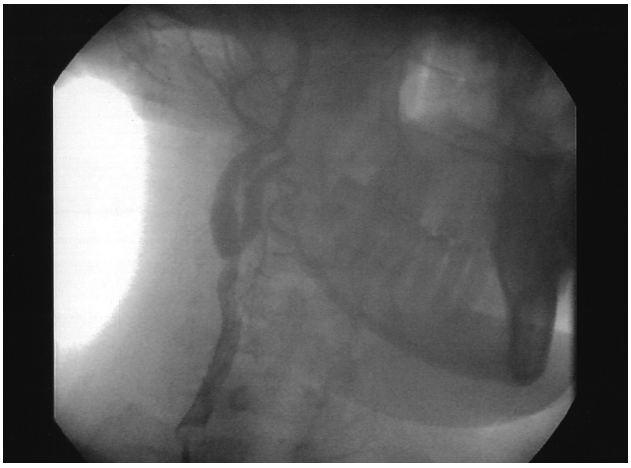


Figure 6. Aortography and angiography demonstrated a plaque in the proximal part of the external carotid artery (ECA) causing 50% obstruction

DISCUSSION

Takayasu's arteritis is a granulomatous giant cell vasculitis which is harmful to the aortic arch and branches. It is known as pulselessness disease or aortic arteritis or aortic arch syndrome (15). Incidence of the disease in North America is predicted to be 2.6/1,000,000 (16). The disease is diagnosed between ages 10-30 in most of the cases and females are affected nine times more frequently. Genetic factors, autoimmunity and various antigens are reported as etiologic factors. Association of TA with systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), CD and ankylosing spondylitis emphasizes the significance of immunity in the pathogenesis of the disease (17).

In CD, association of clinical findings that expose involvement of the other organ systems is well known. In inflammatory bowel diseases (IBD), different clinical findings exist aside from bowel involvement. Among the IBD case reports, it is revealed that nearly all organ systems are affected (18). Uveitis and seronegative spondylarthritis were determined in a case with CD we kept under periodic clinical observation. Clinical association of TA and IBD is rare among literature cases (19). Association of TA and inflammatory disease was investigated in 36 patients retrospectively and chronic or subacute inflammatory disease was diagnosed in 11 of them (20). In Hall et al.'s (21) study with 32 TA patients in North America, CD was diagnosed in two patients. Kerr et al. (22) diagnosed IBD in 4 of 64 TA patients in a wide range study.

High prevalence of the other inflammatory diseases in patients with TA attracts attention to the possible abnormalities of the immunity system (20). Clinical findings and affected organs are almost totally different; however, considerable similarities are present. TA and CD prevalence is high in young females and symptoms frequently occur in the third decade. TA is diagnosed nine times more often in females, indicating the significance of the hormonal effect. CD is a little more frequent in males than females and the hormonal effect is less significant. It was determined in many studies that oral contraceptives increase the frequency of CD; however, clinical findings are incompatible (23-25). It was also reported that hormone replacement therapy-induced CD spontaneously regresses after termination of therapy (26). Our case was a 25-year-old female.

An immunologic mechanism is a common property in both diseases. TH1 lymphocytes are dominant in the lesion. Granulomatous inflammation and increase in gamma-delta cell count in lesions is another common property (27). Histopathological evidence supports the vasculitis hypothesis in the pathogenesis. Ulcers seen in CD characteristically affect the mesenteric face of the bowel. Perivascular granulomas and vasculitis have been determined among biopsies (28,29). Some of the rare symptoms of IBD could be associated with large vessel vasculitis (30).

Constitutional symptoms like fever, weight loss and fatigue may occur during active disease. Ninety percent of the patients suffer from occlusion in the affected vessels. Temporary ischemic attacks, cerebrovascular events, claudicatory upper or lower extremities, angina pectoris, and stomachache are the symptoms caused by ischemia (1). Our patient had fever and stomachache. Difference in blood pressure over 30 mm Hg between arms and murmur over subclavian artery and aorta are the other clinical findings. In our patient, blood pressure was 30 mm Hg lower in left arm.

In the beginning of TA, diagnosis may be delayed due to nonspecific findings. During the inflammation period, high ESR, anemia, leukocytosis, hypoalbuminemia and hypergammaglobulinemia are common findings (17). In our case, high ESR, anemia, hypoalbuminemia and hypergammaglobulinemia were also found. In uncertain cases, diagnosis is confirmed by demonstrating stenosis and aneurysms in the aorta and its main branches with invasive angiography or magnetic resonance

angiography (17). In our study, a 50% stenosis was found in the ECA with aortography and cerebrovascular angiography.

The most differentiating features to distinguish TA from other forms of vasculitis are occurrence of disease before 40 years of age, claudication of extremities, diminished pulse of brachial artery, difference in systemic blood pressure between arms, murmur finding in subclavian arteries and arteriographic findings (17).

Finally, TA should be considered in a young female in the presence of systemic disease and vascular ischemic findings.

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