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Takayasu's arteritis: report of a case with unusual jaw pain

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Abstract

Introduction: Takayasu's arteritis (TA), often referred to as a pulseless disease, is a chronic inflammatory disorder affecting the aorta and its main branches. First reported in 1908 from Japan, it occurs worldwide, but is more prevalent in young oriental females from China and Southeast Asia. The main complications of the disease are due to occlusion of major branches of the aorta. We report a case of an Iranian female who developed left-sided leg intermitent claudication, left arm weakness and left-sided neck and jaw pain.

Keywords: Cardiovascular disorders, Vasculitis, Takayasu's arteritis.

Case Presentation

A 21-year-old woman was referred to the Cardiology Department by otorhinolarynogologistsuspiciousof vasculitis in November 2002. She has experienced gradual and progressive left-lower limb intermittent claudication after 10 meters walking for the last 45 days. She has felt weakness in her left arm activity while combing etc. Left-sided neck pain has also been present for 30 days, and jaw pain on the same side had existed since 8 days before, without tenderness, swelling, inflammation and activity limitation. Adding to her problems. She was also recommended by a denlist for tooth repair, but after two days she felt something like thrilling on the left-side of her neck. After being visited by an otorhinolaryngologist who requested color doppler ultrasonography of the neck vessels she was referred to a cardiologist.At admission, her height was 1.59 cm and she weighed 40 kg.

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Her right radial pulse was normal but it was not palpable in the left side. The blood pressune of her right arm left arm, right and left-leg wene 155/85, undetectable, 160/85 and 120/70 ^{mmHg} respectively. Palpebra was pale but not icteric. Thrilling on the left side of the neck, and to a lesser degree on right side was palpable. Pulmonary examination was unremarkable. No cardiac murmur was audible. The abdomen was soft and flat without tenderness on pressure. Liver, kidney and spleen were not palpable. There was bilateral abdominal bruit around the umblical area. No edema was noticed on legs. Neurological and ophtahalmological examination were normal and fundoscopy showed normal retinal arteries.

Leukocytosis (10000), and a low hemoglobin (10mg/dl) concentration were recorded. Platelet aggregation test revealed no abnormality. A greatly accelerated erythrocyte sedimentation rate of 65 mm/h and an increased C-reactive protein concentration of 3+(no quantitative data) were noted.

Chest x-ray, ECG, and echocardiogram and Arterial color doppler examination revealed increased flow turbulancy of The proximal segment of right subclavian artery, especially at origin (with 60% stenosis), with delayed filling of the distal bed. Color doppler of the left subclavian artery revealed increased systolic flow velocity of the proximal arterial segment with decreased velocity of the diastolic component. All findings were compatible with the 75% stenosis at midportion segment of the left subclavian artery. Intravenous digital subtraction angiography showed significant stenosis at the origin of the right brachiocephalic artery with 60% stenosis at right subclavian artery origin with poststenotic dilatation (Fig. 1) and long segmental stenosis of about 75% and 70% in midportion of left subclavian artery and left carotid artery were seen, respectively, with soft borders (Fig. 2).

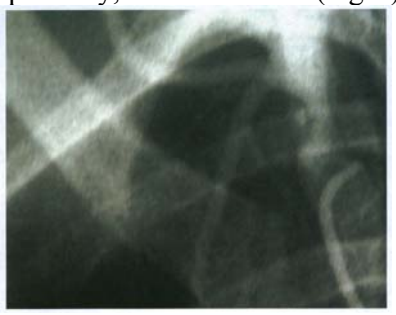

Fig.1: Stenosis at right subclavian artery origin

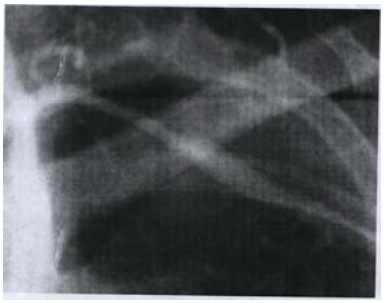

Fig.2: Long segmental stenosis in midportion of left subclavian arterry

In the renal arteries injection, 50% stenosis at origin of both arteries were associated with mild post stenotic dilatation (Fig. 3). Peripheral and coronary arteries were normal.

The patient was diagnosed as having Takayasu's arteritis, and treatment were immediately started with aspirin (100 mg/day), diltiazem (180 mg/day), triamteren-H (50 mg/day), ferrous sulfate (50 mg/day) and prednisolone (50 mg/day).

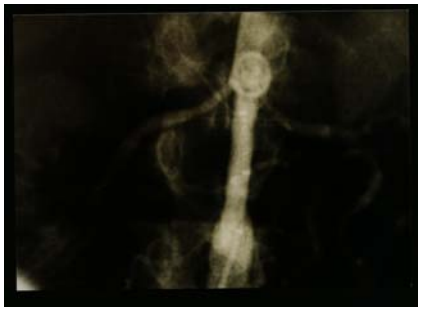

Fig. 3: Stenosis at origin of both renal arteries

Substantial improvement was seen within 5 days. Both ESR and CRP also improved substantially (30mg/dl and 1+, respectively). The patient was discharged on corticosteroid and antihypertensive drugs, and has been well while continuing treatment with prednisolone (5 mg/day) and dilitazem (180 mg/day) for hypertension. Both drugs were continued for 2 months, with further improvement of CRP (Negative) and ESR (25-28mm/h). Regular follow up and discontinuation of prednisolone. Was highly recommended to her during the 2-month follow up no vascular accident occurred.

Discussion

Takayasu's arteritis (TA), often referred to as pulseless disease, is a chronic inflammatory disorder affecting the aorta and its main branches(1). First reported in 1908 from Japan (2), it occurs worldwide, but is more prevalent in young oriental females from China and Southeast Asia (3,4). The main complications of the disease are due to occlusion of major branches of the aorta. The clinical features of absent arterial pulsation in the upper limbs, and head and neck, beside angiographic evidence of brachio-cephalic, left subclavicun, left carotid arterial and renals occlusive disease seen in our patient, are indicative of TA. The extent of her

vascular disease was not established with certainty because our angiographic study was incomplete, and IVDSA lacks precision in detecting arterial lesions clearly and completely. Histopathological examination is required to exclude chronic infective aortitis such as tuberculosis and syphilis in patients with clinical features of TA, and giant cell arteritis in the elderly (4). These conditions would be unlikely in a young female who has no constitutional symptoms and a nonreactive serology for VDRL. Muscle pains, mottled skin, peripheral ischemic events and multifocal encephalopathy which are characteristic of ergotism (5), were not observed in our patient.

Nowadays, doctors who encounter patients with neck pain or inflammatory signs of unknown origin suggested by raised C-reactive protein concentrations and accelerated erythrocyte sedimentation rate would suspect vasculitis and request diagnostic assessment with magnetic resonance angiography, computed tomography, or digital subtraction angiography. Thus the diagnosis of Takayasu's arteritis can by made at an early stage before ischaemic manifestations become obvious (6).

One of the characteristic epidemiological features of Takayasu's arteritis is the preponderance of the disease in young women; in Japan such individuals account for 90% of all patients (7,8). Interestingly, however, an international survey revealed that the female-to-male sex ratio decreases as one moves towards the west (9). This survey also revealed different involvement of the aorta in different countries. Whereas in Japan there is mainly involvement of the ascending aorta, in other Asian countries, the abdominal aorta is more frequently involved, making renovascular hypertension the incorrect diagnosis. TA is considered an immunological disorder based on increased immunoglobulin levels, and is associated with connective tissue disease and endocrine disorder, and with a favorable response to corticosteroid therapy in many patients (1,4). Although there is no reliable index of disease activity in TA, the constitutional symptoms and an ESR of 65 mm in a young female indicate high grade inflammatory disease activity.1 A survey of 859 patients with Takayasu's arteritis in Japan revealed the improvement of their prognosis on the one hand, but an increase in the potential complications of atherosclerotic disease on the other, because inflammation is a serious risk factor for atherosclerosis (10).

The patient presented in Japan carried the HLA A24-B52-DR2 haplotype, which is closely associated with Takayasu's arteritis (11). Patients with this haplotype are prone to accelerated inflammation more than patients without this haplotype. This patient could have initially had an infection, such as tooth decay, alveolar pyorrhoea, or the common cold, which may then have induced acute vasculitis through activation of autoimmune mechanisms. A genetic predisposition to the development of TA is suggested by the occurrence of familial cases (4,12), but in our case, there is no familial history. In this case, cardiac auscultation revealed no cardiac murmur. It is essential, particularly in Japan, to check whether aortic regurgitation due to dilatation of the aortic root is present, because this complication is very frequent (9,13).

Aortic regurgitation can induce congestive heart failure or arrhythmia, both of which are the main causes of death.9 However, patients with Takayasu's arteritis in other Asian countries such as China,(14) Thailand,(15) and India(16) have a high incidence of cerebral accidents due to renovascular hypertension, which is the main cause of death. There is not published literature in this field in Iran. A standard medical treatment for Takayasu's arteritis in Japan is the use of antiplatelet drugs (low dose of aspirin, for instance) and steroids as anti-inflammatory therapy.

Immunosuppressive drugs are used only as supplementary therapy because of their adverse effects (6). Surgical intervention is sometimes of value to prevent ischemic complications, but results are less favorable in patients with advanced disease(1,4). The eight-week period of observation was too short to assess treatment effects. However, the blood pressure in the upper limbs had improved slightly. The estimated median duration of corticosteriod therapy before remission in TA is about 22months (17). Regression of vascular stenotic lesions has been reported in steroid-treated TA patients(18). Long-term survival in TA patients is better than 80% after 20 years of follow-up.1,4 A high index of suspicion is therefore required for the diagnosis of TA at the presymptomatic phase 1 stage, when treatment may induce remission and complications may be avoided or minimized.

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خلاصه

آرتریت تاکایوسو: گزارش یک مورد غیر معمول از درد فک

دکتر افسون فضلی نژاد، دکتر مهدی بخشایی، دکتر نوید نوری زاده

مقدمه: آرتریت تاکایوسو(TA)، یک اختلال مزمن التهابی است که عروق بزرگ قبل آئورت و شاخه های اصلی آن را درگیر می کند و به عنوان بیماری بدون نبض شناخته می شود. این مورد اولین بار در سال ۱۹۰۸ از ژاپن گزارش شد و در تمام دنیا گسترش دارد اما در خانم های جوان شرقی در چین و آسیای جنوب شرقی شایعتر است. عارضه اصلی بیماری، انسداد شاخه های اصلی آئورت می باشد، گزارش ، شامل یک خانم ایرانی که با لنگش متناوب پای چپ و ضعف بازوی چپ همراه با درد متناوب گردن و فک در سمت چپ مراجعه کرده بود.

واژه های کلیدی : اختلالات قلبی–عروقی، واسکولیت، آرتریت تاکایاسو