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Case Report
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Unforgettable Moments in Hypertension a Rare Case of Takayasu Disease

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Abstract

This report describes a unique case of a young Middle Eastern female presented with 3 - 4 years history of non specific complaints including headache, fatigue, dizziness, poor appetite, irregular menses, sense of suffocation, weight loss and transient right knee joint painful swelling. On examination, she was found to have high BP measurement at right upper limb with low BP measurement at left upper limb, weak left radial pulse with left subclavian and abdominal bruits. Computed Tomography angiographic examination of the aorta showed multiple lesions at aorta and its major branches ranging from aneurysmal dilatation through attenuation, stenosis into total occlusion with diagnosis of Takayasu arteritis. She responded well to treatment with steroid and renal artery stenting.

Keywords: Takayasu arteritis; Hypertension; Renal artery stenosis; Subclavian artery occlusion

Introduction

Takayasu arteritis is a chronic vasculitis of unknown etiology [1]. Women are affected in 80 to 90 percent of cases, with an age of onset that is usually between 10 and 40 years [1,2]. It has a worldwide distribution, with the greatest prevalence in Asians [3-6]. In Japan, it has been estimated that 150 new cases occur each year [7]; the incidence is one to three new cases per year per million population in the United States and Europe [2]. Human leukocyte antigen (HLA)-Bw52 and HLA-B39.2 have been increased in frequency in several studies, suggesting an immuno genetic association [8]. We report an interesting case of a young female with multiple complaints and hypertension with significant discrepancy and gradient between her right and left upper limb blood pressure measurement. Multi slice Computed Tomography (CT) angiographic examination of the aorta showed saccular aneurysmal dilatation of the upper abdominal aorta, markedly attenuated proximal segment of superior mesenteric artery with significant collaterals from inferior mesenteric artery, markedly stenotic proximal segment of right renal artery and early occlusion of left subclavian artery with provisional diagnosis of Takayasu arteritis.

Case Presentation

This is a 23 year old Middle Eastern female studying abroad presented to multiple medical services with few years history of non specific multiple complaints including headache , dizziness , general weakness, lethargy, easy fatigability, irregular menses, dizziness, arthralgia and poor appetite. No dyspnoea, chest pain, neurological or visual complaints. Her initial physical examination showed BP measurement at left upper limb of 100/60 mm Hg and was diagnosed as anxiety depression and anxiolytics were prescribed.

Later on, patient continued to experience same complaints with weight loss, sense of suffocation and swollen painful right knee joint. She denied any other complaints. She is non smoker with negative drug, surgical and family histories. Her physical examination revealed underweight girl with BMI 16.4, BP measurement of 160/95 mm Hg in right upper limb and 95/58 mm Hg in left upper limb. Her CVS examination showed left weak radial pulse with left subclavian and abdominal bruits with no cardiac murmurs and clear chest, and otherwise normal abdominal, neurological and skin exam with swollen non tender right knee joint.

Investigations showed mild hypochromic microcytic anaemia, normal renal and liver functions; normal fasting blood sugar and TSH with normal lipid profile. ECG and CXR were within normal limits. Echocardiography showed normal LV systolic function with EF 65%, mild left ventricular hypertrophy and no valvular lesions. Erythrocyte sedimentation rate was 32 and also CRP level was slightly elevated 24 mg/l.

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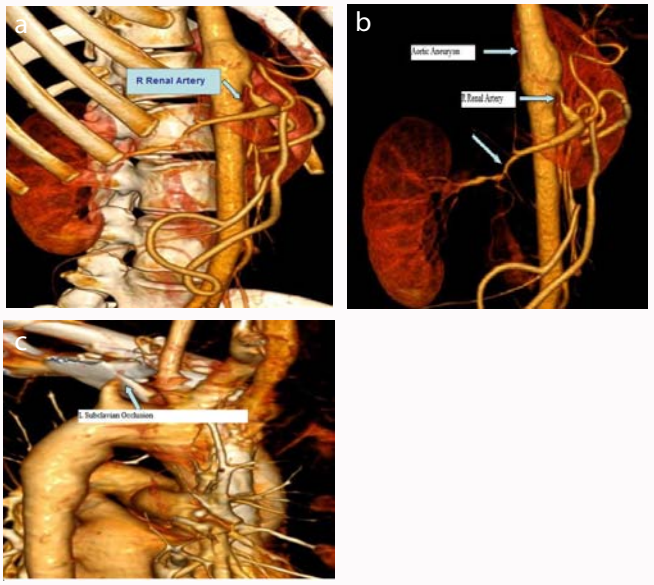

Figure 1:

Multi slice Computed Tomography angiographic examination of the aorta showed mild saccular aneurysmal dilatation of the upper abdominal aorta above the celiac artery, markedly attenuated proximal segment of superior mesenteric artery with significant collaterals from inferior mesenteric artery, markedly attenuated proximal segment of right renal artery and early occlusion of left subclavian artery (Figure 1a-c). Patient was evaluated by ophthalmologist, neurologist and interventional cardiologist and assessment of retinal, coronaries, and cerebral circulation were done. Prednisolone and baby aspirin 81mg daily were added to her medication by rheumatologist to overcome disease activity.

Blood pressure continued to be high after 4 weeks of treatment, so nifedipin LA 30 mg daily was added. Patient had better control of BP. Renal artery stenting was considered but postponed until complete remission of activit According to criteria of the American College of Rheumatologists [2] and to modified criteria for diagnosis of Takayasu Disease [2-6], patient has satisfied criteria for diagnosis of Takayasu Disease, and it is considered late phase of the disease with type V involvement. Patient was followed up and later had undergone right renal artery stenting with no complaints and controlled blood pressure.

Discussion

Takayasu arteritis is a chronic vasculitis of unknown etiology [1]. Women are affected in 80 to 90 percent of cases, with an age of onset that is usually between 10 and 40 years [1,2]. It has a worldwide distribution, with the greatest prevalence in Asians [3-6]. In Japan, it has been estimated that 150 new cases occur each year [7]; the incidence is one to three new cases per year per million population in the United States and Europe [2]. Human leukocyte antigen (HLA)-Bw52 and HLA-B39.2 have been increased in frequency in several studies, suggesting an immunogenetic association [8]. Takayasu arteritis primarily affects the aorta and its primary branches [9]. The inflammation may be localized to a portion of the thoracic or abdominal aorta and branches, or may involve the entire vessel. Although there is considerable variability in disease expression (due perhaps to geographic differences [9], the initial vascular lesions frequently occur in the left middle or proximal subclavian artery. As the disease progresses, the left common carotid, vertebral, brachiocephalic, right middle or proximal subclavian artery, right carotid, vertebral arteries, and aorta may also be affected. The abdominal aorta and pulmonary arteries are involved in approximately 50 percent of patients.

The inflammatory processes cause thickening of the walls of the affected arteries. The proximal aorta may become dilated secondary to inflammatory injury. Narrowing, occlusion, or dilation of involved portions of the arteries in varying degrees results in a wide variety of symptoms [10] our case is an unusual presentation of a rare disease. It is very important to perform full physical examination to any patient even if his / her presentation is very non specific complaints. Also, it is mandatory to check BP in both arms at least on first presentation, and to compare pulses at all limbs and to evaluate more if any abnormality. The importance of early diagnosis is of great help as it might delay or prevent any progression of the disease.

Diagnosis of Takayasu disease depends on suspicion and presence of specific major and / or minor criteria. Multidisciplinary medical care is of paramount importance in managing such patients in order to evaluate all potential organs.

The best recommended treatment to stop disease activity is prednisolone especially if it is given early enough and to switch into immunosuppressive drugs like methotrexate etc. when prednisolone is ineffective. Addition of aspirin might be needed if thrombosis is suspected or expected [11] Interventional treatment is most rewarding when stenting of arterial stenosis is indicated. Coronary artery bypass grafting using venous conduits is superior to stenting of affected coronary arteries even if DES is utilized. Unfortunately, delay in diagnosis is very common even if patient presents to a vascular specialized physician.

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