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Case Report

Vasculitis masquerading as a mass: a case report of Takayasu arteritis in a 28-year-old male

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ABSTRACT

Takayasu arteritis is a chronic vasculitis mainly involving the aorta and its main branches most commonly subclavian and carotid. It induces clinically varied ischaemic symptoms due to stenotic lesions or thrombus formation, including blindness, retinal haemorrhage, pulselessness, aortic regurgitation and congestive heart failure due to dilatation of the ascending aorta. More acute progression causes destruction of arterial wall, leading to the formation of aneurysms and rupture of the involved arteries. Here we present a case of Takayasu's arteritis in a 28-year-old male who presented with nonspecific symptoms of fever and neck pain with palpitations and feeling of pulsations in right side of neck since last 20 days and was eventually diagnosed as arteritis. This case shows that it can present with many non-specific symptoms and can be diagnosed with proper examination and a high index of suspicion due to its nonspecific overlapping features with many diseases.

Keywords: Panarteritis, Pulselessness, Aortic regurgitation, Aneurysm

INTRODUCTION

Takayasu arteritis is also known as pulse-less disease. It leads to inflammation of the large and medium arteries with their branches. It presents with mononuclear and granulomatous changes. This leads to transmural fibrous thickening of the walls of arteries. This further results in several vascular occlusions and/or aneurysmal dilatation which may lead to ischemic changes and other constitutional symptoms like headache, fever, unalike blood pressure, syncopal attacks.

Takayasu predominantly affects young Asian women. A systematic review of the literature has demonstrated incidence rate of Takayasu is 1.11 (95% CI 0.70-1.76) cases per million person-years.[1] However, our patient who was a young male presented atypically with a pulsatile neck mass without any symptoms of limb claudication, syncope and pulselessness making it difficult to reach the diagnosis.

CASE REPORT

A case of a 28-year-old male, presented with fever for 20 days and pain in the neck region from last 18 days. Fever was insidious onset, low grade, and associated with palpitations, headache and chest discomfort. Pain in right side of neck started 18 days back associated with pulsatile feeling in that region. No history of hypertension, neck trauma, Tuberculosis contact, syncopal attacks. Patient didn't experience similar symptoms in the past and none of the family members had similar complaints. Patient had smoking index of 30 and is an occasional drinker. Systolic blood pressure was 142 mm of Hg in right upper limb, and 138 mm of Hg in left upper limb, 184 mm of Hg in right lower limb with 182 mm of Hg recorded in left lower limb. Oxygen saturation was 98% on room air, pulse rate was 82 bpm with characteristic water hammer pulse and no sign of elevated JVP. Head to toe examination revealed swelling in right side of neck approximately 2×1 cm which was

erythematous, pulsatile with raised local temperature. CVS examination revealed apex beat in 6th IC space 3 cm lateral to midclavicular line and a decrescendo blowing diastolic murmur in the aortic area radiating to apex. CBC revealed WBC count of 45.25 thousand and Hb 9.4 gm/dl. Inflammatory markers were raised including ESR of 120 mm in 1st hour with positive CRP. USG neck revealed abnormal dilation of right CCA with swirling of blood flow in color doppler along with wall thickening in left CCA extending to ICA. Blood cultures were sent and 2D Echocardiography was performed revealing dilated aortic root and dilated ascending aorta with moderate AR. No evidence of clot or vegetation was seen. CT angiogram of neck showed soft plaque in right CCA with luminal narrowing at origin and the rest of CCA and proximal ICA are dilated suggestive of aneurysm. Mild dilation of left ICA giving beaded appearance p/o vasculitis.

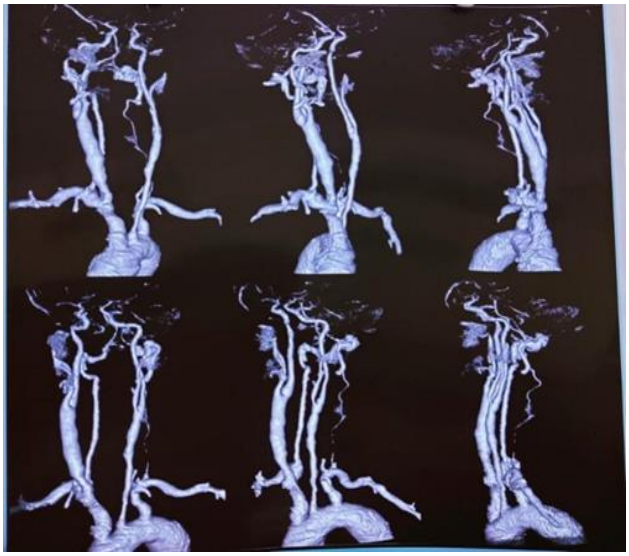

Figure 1: CT angiogram of neck.

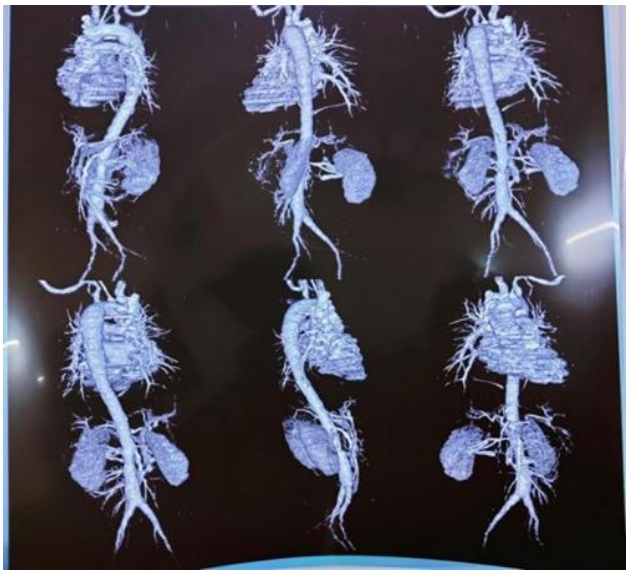

Figure 2: CT aortogram.

CT aortogram revealed mild aneurysmal dilation of ascending and arch of aorta. Segmental narrowing with distal dilation was seen in abdominal aorta and bilateral common iliac arteries with thickened irregular walls, suggestive of arteritis. According to the study by Ishikawa, criteria to clinically diagnose Takayasu (sensitivity of 84%) consists of one obligatory criteria (age less than or equal to 40 years); two major criteria (left and right mid subclavian artery lesions); and nine minor criteria including high ESR, hypertension, CCA tenderness, AR, lesion in pulmonary artery, left CCA, distal brachiocephalic, thoracic and abdominal aorta.[2] Diagnosis requires 2 major criteria or 1 major + 2 minor criteria or 4 or more minor criteria along with obligatory criteria of age. Our patient fulfilled six minor criteria along with obligatory criteria of age namely high ESR, hypertension, CCA tenderness, AR, thoracic and abdominal aorta lesion. So clinical diagnosis of Takayasu arteritis was made. Blood culture and urine examination were negative. Following therapy with IV antibiotics, TLC of the patient normalized after which he was started on oral prednisolone and methotrexate, which improved his symptoms.

On follow up visit after 2 weeks, patient reported clinical improvement and continued to be asymptomatic. Prednisolone was continued with tapering dose for further remission

DISCUSSION

Takayasu arteritis is the most common cause of renovascular hypertension in India, China, Japan, Korea and other south east Asian countries. At its core, it is an inflammatory disease, and it is thought that autoimmune cell-mediated immunity may be responsible for the disease. But its etiology is still largely unknown. Diagnosis is based on suspicion as well arteriographic findings. There continues to be a substantial lack of understanding of the pathogenesis of Takayasu arteritis however Involvement of CD4+ and CD8+ T cells may play a key role in the pathophysiology of Takayasu arteritis, as these cells support the formation of granulomas and potentially activate the activities of various proteases such as matrix metalloproteinase (MMP), as well as other cells which promote chronic inflammation and fibrosis formation. There has also been speculation about a potential link with certain human leukocyte antigens (HLA). The strongest association has been established with HLA-B52 in Japanese and other populations.[3,4]

Histology encompasses a spectrum ranging from adventitial mononuclear infiltrates and perivascular cuffing of vasa vasorum to intense transmural mononuclear inflammation, to granulomatous inflammation full of giant cells and patchy medial necrosis. The inflammation is associated with irregular thickening of vessel wall, intimal hyperplasia and adventitial fibrosis.

Many studies have reported a female to male ratio of 1.58:1 in Indian patients. Indian male patients tend to have involvement of abdominal aorta with hypertension.[5] The average age of presentation is in the third decade. The incidence of aortic regurgitation has been low (7-15%) in many series from India.[6]

The initial signs and symptoms are nonspecific including fatigue, weight loss and fever mimicking diseases like tuberculosis. On progression, vascular signs and symptoms appear that include reduced upper extremity blood pressure and pulse strength clinically seen as arm claudication and Raynaud's phenomenon; neurological deficits including syncope, transient ischemic attacks, stroke and ocular disturbances. These symptoms depend on the vessel involved.

Differentials to our case includes atherosclerosis, fibromuscular dysplasia, tuberculosis and giant cell arteritis. Evidence based exclusion of the former differentials as well as the radiological and examination findings led us to the diagnosis of Takayasu.

CONCLUSION

This case contributes to the expanding comprehension of the Takayasu arteritis highlighting its clinical aspects and developing avenues for appropriate diagnostic and imaging modalities as-well-as intervention techniques. The motives of this article extend beyond the clinical case, influencing future research, diagnostic and prevention strategies.

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