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Case Report

A Case Report: An Unusual Presentation of Takayasus Arteritis

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Abstract

Takayasu's arteritis (TA) is a chronic granulomatous vasculitis of unknown cause. It is characterized by stenosis, occlusion and aneurysm of large elastic arteries. Commonly arch of aorta and its branches. It mainly affects young women of Asia, Middle East and South America. The presented report is the case of a young woman of 18 years age from Faridpur, Bangladesh having the atypical and uncommon features of Takayasu's arteritis. Her complaints were fever, weakness of left hand, pain over back of chest and neck and dizziness with vomiting for several times. The report also covered details of case history, selected investigations, and treatment modalities including author's views in terms of discussion and conclusion in particular.

Introduction

Takayasu's arteritis (also known as, "aortic arch syndrome," "nonspecific aortoarteritis," and "pulseless disease") is a form of large vessel granulomatous vasculitis with massive intimal fibrosis and vascular narrowing, most commonly affecting young or middle-age women of Asian descent, though anyone can be affected[1]. It mainly affects the aorta (the main blood vessel leaving the heart) and its branches, as well as the pulmonary arteries. Females are about 8–9 times more likely to be affected than males. There are various clinical presentations of TA. Some time patient's presentations are nonspecific. This particular patient presented in an uncommon way.

Case History

A female patient 18 years of old, normotensive, non-diabetic, non-smoker, hailing from Digholia, Modhukhali, Faridpur got admitted in Diabetic Association Medical College Hospital on 31.03.2019 at 1.30 PM presented with low grade fever for one and half months, it was intermittent in nature, starts at evening without chill and rigor. It then subsided with sweating and after taking Paracetamol at night. She had no cough, chest pain, Haemoptysis, and had significant weight loss associated with fever. She also had weakness of left hand associated with claudication pain on raising left hand over 10 to 15 minutes of manual work and that subsided after taking rest. For last 1 month she had complains of pain on the back of chest and neck without any radiation on flexion of neck. She also complained for dizziness for last 15 days associated with vomiting for several times. Dizziness increased on rising on sitting from supine position associated with feeling of darkness which relieved by lying flat.

She was diagnosed as a case of rheumatic fever 10 years ago and treated with PHENOXYMETHYL PENICILLIN but there is no clinical evidence for that diagnosis. She admitted in this hospital 3 months ago and diagnosed as Urinary Tract Infection and treated with NITROFURANTOIN. She is a college student. She got married for 2 years with sedentary life style. She had no history of smoking, betel nut or tobacco leaf or gul intake. No one of her family member was suffering from such type of disorder. She was poor, drinks tube well water and uses sanitary latrine, lives in a tin shaded house with ill ventilation. She was immunized as per EPI schedule .There was no history of recent travelling to hill track area or north Bengal. Her menstrual cycle was regular with normal duration and flow. She has leucorrhea and valval itching.

Patient was severely anemic and mildly emaciated. Her pulse and blood pressure was not recordable on the left arm and hand. But on the right upper limb B.P. was 120/80 mm of Hg and pulse 88 beat per minute & regular. Carotid bruit was present over both right and left common carotid artery and also left sub-clavian artery. Movement of left shoulder was restricted not above the shoulder, and was painful. Examination of other systems revealed no abnormality.

Her laboratory investigation revealed Hb-8 gm/dl, ESR- 85 mm in 1st hour, TC of WBC- 12.67^3/ul, Polymorph 68%. C-Reactive Protein (CRP) was normal. Duplex color doppler study of arch of aorta revealed stenosis of left common carotid artery, left subclavian artery and right common

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carotid artery. Findings were consistent with Takayasu's Arteritis. Patient received prednisolone and methotrexate on diagnosis. After one week she was much improved.

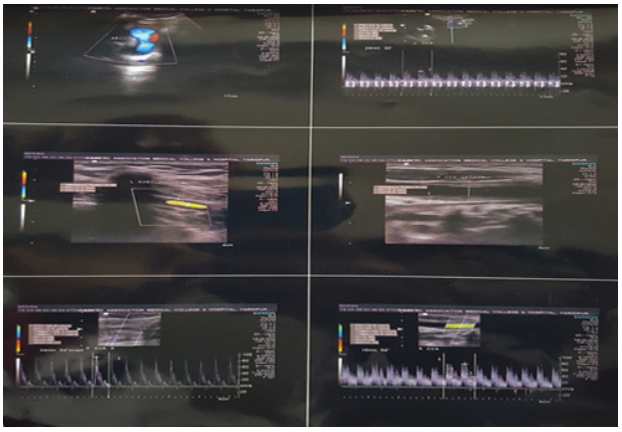

Figure1: Duplex colour doppler study of arch of aorta and its branches shows significant narrowing of RCCA, LCCA, Left subclavian artery consistent with Takayasu's arteritis.

Discussion

Takayusu's arteritis has been reported from all over the world, there is a wide variation in its prevalence different countries. It is predominant in Asia, Middle East and South American countries. The incidence rate of Takayusu's arteritis in Japan is 4.2 persons per 100000 populations. Takayusu's arteritis affects healthy women, the average age of onset in Japanese and Indian patient being 15-35 years. TA is a form of large vessel granulomatous vasculitis[2-4] with massive intimal fibrosis and vascular narrowing.

Although the cause of Takayasu's arteritis is unknown, the condition is characterized by segmental and patchy granulomatous inflammation of the aorta and its major derivative branches. This inflammation leads to arterial stenosis, thrombosis, and aneurysms.[3] There is irregular fibrosis of the blood vessels due to chronic vasculitis, leading to sometimes massive intimal fibrosis (fibrosis of the inner section of the blood vessels). [4,5]

In the Western world, atherosclerosis is a more frequent cause of obstruction of the aortic arch vessels than TA. TA is similar to other forms of vasculitis, including giant cell arteritis which typically affects older individuals.[2] Due to obstruction of the main branches of the aorta, including the left common carotid artery, the brachiocephalic artery, and the left subclavian artery, Takayasu's arteritis can present as pulseless upper extremities (arms, hands, and wrists with weak or absent pulses).[6,7]

The genetic contribution to the pathogenesis of Takayasu's arteritis is supported by the genetic association with HLA B*52.[8] In addition, a genetic association was identified and confirmed between Takayasu's arteritis and the FCGR2A/FCGR3A locus on chromosome 1 (rs10919543, OR=1.81, p=5.89×10-12). The risk allele at this locus results in increased mRNA expression of FCGR2A. A genetic association between IL12B and Takayasu's arteritis was established.[9]

Patients are usually presented with acute and chronic symptoms. In the acute stage the symptoms of Takayasu's arteritis are usually nonspecific and generalized, including fever, easy fatigability, general malaise, neck pain, weight loss, and arthralgia. Faintness and/or dizziness are sometimes reported, perhaps due to hypersensitivity of the baroreceptors in the aortic arch leading to hypotension.

In the chronic stage most patients with Takayasu's arteritis present with features related to specific vascular lesions, although they may also have the constitutional symptoms described above. In East Asian countries the condition affects the aortic arch most frequently, hence typical complaints relate to ischaemia of the brain, eyes, or arms, namely dizziness, syncope, visual disturbance, and easy fatigability of the arms, with pain due to intermittent claudication.

Occasionally TA has some atypical presentation. systemic illness with signs and symptoms of malaise, fever, night sweats, weight loss, joint pain, fatigue, and fainting. Fainting may result from subclavian steal syndrome or carotid sinus hypersensitivity.[4] There is also often anemia and marked elevation of the ESR or C-reactive protein (nonspecific markers of inflammation). The initial "inflammatory phase" is often followed by a secondary "pulseless phase".[2] The "pulseless phase" is characterized by vascular insufficiency from intimal narrowing of the vessels manifesting as arm or leg claudication, renal artery stenosis causing hypertension, and neurological manifestations due to decreased blood flow to the brain.[10]

This particular patient presented in the same atypical way, thus consulted with different specialist. After diagnosing her as a case of Takayasu's arteritis and treating her with Prednisolone and Methotrexate a dramatical response was found.

Conclusion

Some time in case of Takayasu's Arteritis early diagnosis is difficult especially when presented atypically. Therefore, careful history taking and clinical examination should be considered important towards a diagnosis.

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