CASE REPORT

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A Case of Takayasu Arteritis in a Male Patient

Suvam Banerjee[1], Elina Semenenko[2]

1. Department of Health and Family Welfare, Burdwan Medical College and Hospital (WBUHS), Government of West Bengal, India;
2. Faculty of Psychology and Neuroscience, Maastricht University, Netherlands*

ABSTRACT

Takayasu arteritis (TA), otherwise called "pulseless disease", is a category of primary systemic vasculitis involving medium and large arteries, including the aorta, its branches and the pulmonary and the coronary arteries. The presence of TA is often observed in women and the main population of patients develops their first symptoms between 15 years to 30 years. If left untreated, this disease may lead to the development of many comorbid renal and pulmonary obstructions causing lethal consequences.

Here we present a case report of the diagnosis of TA in a 25-year-old male, treatment and following procedures. A patient with no anamnesis was hospitalized with paralysis of the right half of body and altered consciousness. He had been experiencing symptoms like fatigue, general weakness and respiratory distress for the past year. After extensive medical analysis patient was treated with oral prednisone and later endarterectomy was performed. After the operation, the patient improved and was discharged. Till date, no new complaints were reported.

Keywords: Takayasu Arteritis, Primary Systemic Vasculitis, Aneurysm, Rheumatology

*See End Note for complete author details

INTRODUCTION

TA is a form of inflammatory large vessel granulomatous vasculitis of unknown etiology, most often affecting medium and large arteries.[1] It is a rare disease, with a worldwide prevalence of approximately 1 to 3 new cases per million occurring every year, majority of patients are females, yet cases of TA were recorded in children and elderly people as well.[2] Most TA cases are recorded in Asian countries, even though this disease is recognized worldwide.[2] The most common consequences are thickening of tunica intima of blood vessels, claudication of extremities, irregular fibrosis, thrombosis and stenosis.[3] Due to stenosis of the aorta or pulmonary arteries, patients diagnosed with TA can exhibit feeble or no pulse in upper extremities (arms, hands, wrists).[1] All mentioned symptoms can lead to different pathological complications including ischemia in different organs, aneurysm or dilatation of the associated vessels.

CASE REPORT

A 25-year-old male, resident of West Bengal, India, with no known history of comorbidities was introduced to the medical clinic, on 3rd January 2018, with insidious onset of aphasia and right-sided weakness for eight days. As indicated by the patient's friend, he was apparently well eight days back when he developed an altered level of consciousness, which drove them to carry him to the clinic. In the medical clinic, the patient regained consciousness on the admission day, however, he could neither talk nor move the right half of his body. There was no history of fever, fits, nausea, vomiting, heaving, urinary incontinence, fecal incontinence, as well as any visual disturbances. This helped rule out certain clinical disorders affecting the nervous system of our body which might lead to raised intracranial pressure. These differentials include meningitis, encephalitis, meningoencephalitis, epilepsy, febrile seizures, metabolic disorders with dyselectrolytemias, poisoning, infections with chance of sepsis. However, the patient had complained of malaise, generalized weakness, respiratory distress and fatigue for the last year.

Additionally, the patient also complained of unilateral headache which was not relieved by over-the counter NSAIDS. The patient had no other relevant past medical, surgical or family history. On arrival, his blood pressure was 150/96 in the left brachial artery, however non-recordable in

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Corresponding author:

Dr. Suvam Banerjee MBBS, Clinical Intern, Department of Health and Family Welfare, Burdwan Medical College and Hospital (WBUHS), Government of West Bengal, India.
E-mail: banerjeesuvam1@gmail.com Mobile: +918777659706

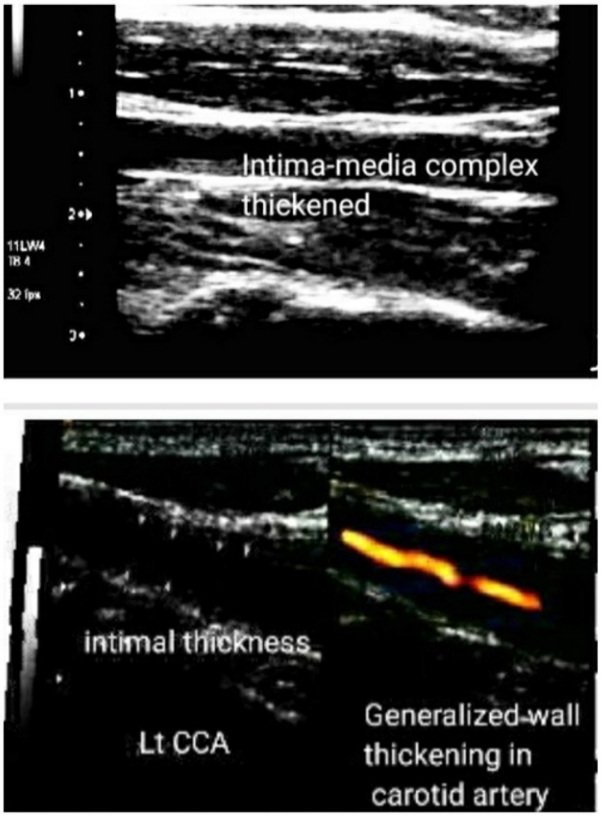

Figure 1. There is thickening of the intima-media complexes in the wall of the blood vessels in case of Takayasu arteritis. In our case report, we detected thickened intima of common carotid artery (CCA) with generalized wall thickening of carotid artery

the right arm, his pulse was 60 beats/minute (just recordable in the left radial artery), his temperature was 36.7°C and his respiratory rate was 22 breaths/minute. On assessment, his overall physical, respiratory and abdominal examinations were normal. However, a central nervous system assessment revealed problems, with aphasia in speech. Motor system assessment on the right side revealed decreased power in both upper and lower limbs as well as upgoing planters on the right lower limb. The rest of the assessment was normal. His cardiovascular assessment showed muffled heart sounds,

Table 1. The American College of Rheumatology criteria for classification of Takayasu Arteritis

Sl. No.	Criteria
1	Age at onset of disease <= 40 years
2	Extremities of the body experiencing claudication
3	Reduced brachial artery pulse
4	Blood pressure difference >10 mm Hg
5	Bruit over subclavian arteries or aorta
6	abnormalities in arteriogram

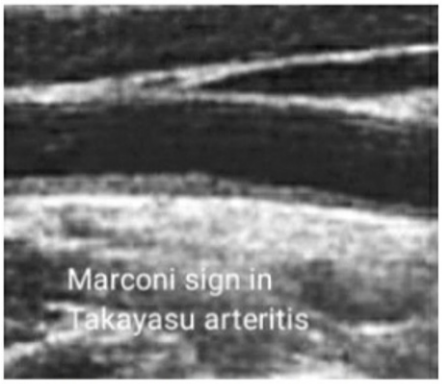

Figure 2. An image showing "Marconi sign" found in Takayasu arteritis

and murmurs were absent, yet he was positive for carotid bruit on both sides.

Based on the history and examination, we ordered appropriate laboratory tests along with other specific tests. When his baseline lab values were ordered, complete blood count (CBC) showed hemoglobin (Hb) of 12.6 g/dL (typical: 11.1-14.5 g/dL), mean corpuscular volume of 77.8 fL (ordinary, 76-96 fL), WBC count of 8.5×10^9/L (typical: 4-10×10^9/L) and platelets were 200×10^9/L (ordinary, 150-400×10^9/L); however, C- reactive protein was 180 mg/L (ordinary <3 mg/L) and erythrocyte sedimentation rate was 90 mm/hour (typical, 0-22 mm/hour). As the patient had intermittent respiratory distress, we ordered a chest x-ray and echocardiography, both of which did not show any abnormality. The patient's coronary and renal angiograms were also normal.

As the carotid bruit was audible on both sides, we also performed carotid color Doppler imaging (assessing the common carotid artery (CCA), internal carotid artery (ICA), external carotid artery (ECA) and vertebral artery (VA)). Findings showed diffuse homogenous intimal thickness involving the bilateral CCA, ICA, and ECA **(Figure 1),** exhibiting macaroni signs **(Figure 2)**, causing significant luminal constriction. Other findings include minimal flow and decreased peak systolic velocity (PSV) in the right CCA and ICA on power Doppler and no flow on color and power Doppler images in the right ECA. The right VA was dilated with normal flow and no flow was seen in the PSV, left proximal and mid-CCA on power Doppler images (suggestive of complete occlusion). However, the left ICA and ECA showed reduced flow and diminished PSV on power Doppler (due to collateral supply), the left VA shows no flow (suggestive of complete occlusion) and there was turbulent flow in the aortic arch.

By and large, based on clinical manifestations and carotid artery Doppler findings, a diagnosis of TA was made. We

began the patient on oral prednisone (60 mg) once daily on a tapered basis and consulted the department of vascular surgery for potential surgical intervention. The patient had an endarterectomy fourteen days after admission which caused an improvement in his symptoms, after which he was released on 5th February 2018, and no follow-up has since been needed.

DISCUSSION

The typical patient with diagnosed TA will be a young woman of Asian descent in her second or third decade of life. Even though this disease occurs worldwide, TA is most commonly affecting women of Asian descent, making cases of TA in women happen 8-9 times more than in men.[4] This case study was of particular interest to the authors, as the patient diagnosed with TA was an Indian male with no known comorbidities and no anamnesis of health issues. According to the American College of Rheumatology criteria **[Table 1]** for clinical diagnosis of TA, the patient presented in this report met 3 criteria out of 6, which was necessary to fulfil the criteria for diagnosis.[4] Criteria include the age of disease onset < 40 years, claudication of extremities and arteriogram abnormalities. Treatment for TA should include medical therapy with steroids to which half of the patients respond.[5] The other half of the patients are given immunosuppressants.[5] The steroids and immunosuppressants have the following mechanism of action- they decrease the inflammation and suppress the activity of the immune system. Since the inflammatory mediators with hyperactive immune system leads to the causation and manifestations of TA, which is most likely an autoimmune disease, these drugs become highly effective as they suppress this and help reduce the impact of the autoimmune disease on the body.[6] Surgical interventions are also required sometimes. However, they have a high failure rate, especially if surgeries are performed during the inflammatory acute stage. Nowadays, PET scans have significant role in assessing whether the arteritis is active or not. They can detect the active stage not only in patients with active disease before treatment but also in relapsed cases receiving steroids and immunosuppressive drugs.[7]

The major cause of concern is the lack of awareness of patients about TA which leads to late presentation and this delay in diagnosis causes complications.[8] To improve that and to promote admission to hospital promptly, awareness promotion and regular screening of the patients by professionals to exclude comorbidities or complications should be carried out.

END NOTE

Author Information

1. Suvam Banerjee MBBS, Clinical Intern, Department of Health and Family Welfare, Burdwan Medical College and Hospital (WBUHS), Government of West Bengal, India.
2. Dr. Elina Semenenko MS, Neuro-psychology, Intern and Research Assistant, Faculty of Psychology and Neuroscience, Maastricht University, Netherlands

Conflict of Interest: None declared

Participant Consent: Written informed consent for publication of the clinical details was obtained from the patient. As informed consent was taken from patient, the patient was taken into confidence that the data will only be used solely for research purpose, and his confidentiality was maintained and no medical intervention was done.

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