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CASE REPORT

Asymptomatic Patient with Unrecordable Blood Pressure: Could It be Takayasu?

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ABSTRACT

Takayasu's arteritis is a rare, systemic inflammatory vasculitis of large vessels that usually affects women of childbearing age. Also known as pulseless disease or occlusive thromboaortopathy, the disease is named after the Japanese ophthalmologist who in 1905 described a form of retinal arteriovenous anastomoses due to retinal ischemia caused by large vessel vasculitis. We present here a case of a young female who presented with stroke. The patient had stroke which eventually recovered but now was referred for evaluation of her unrecordable blood pressure.

Keywords: Blood pressure, Stroke, Takayasu's arteritis.

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CASE PRESENTATION

A 24-year-old Hindu female patient was referred to our department for low blood pressure (BP). She was asymptomatic at present. She gave history of a left-sided hemiparesis with slurred speech 2 years earlier for which she was hospitalized under care of a neurophysician in a charitable hospital. Computed tomography (CT) brain was carried out, she was treated with aspirin and clopidogrel. She recovered in 3 weeks period except slight blurring of vision. She was following up with neuro outpatient department for past 2 years. A house physician referred her to a medicine outpatient with a note of low BP. Her discharge summary did not mention her pulse or BP during her hospital stay or subsequent follow-up. It only mentioned left-sided hemiparesis with left upper motor neuron facial paralysis, with noncontrast CT scan of brain suggestive of acute infarct in right parietal lobe. A residual neurodeficit in the form of minimal weakness of left hand persisted.

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She is not a known hypertensive or diabetic. There is no history of sensory complaints or any arm or leg claudication. The family history is not suggestive of early-onset cerebrovascular or connective tissue disorder, hypothyroidism, or any other illness. Presently, she complained of minimal blurring of vision for which the resident doctor in neuro outpatient department checked her BP and then referred the case to us. In our outpatient clinic it was found that her bilateral radial, brachial, carotid pulsations were absent. Right femoral pulse was weakly palpable and both dorsalis pedis pulsations could be felt. The BP obviously could not be measured. No bruit was heard over any peripheral artery. The systemic examination was within normal limits except slightly weak grip of the left hand. A case of young hemiplegic female with absent peripheral pulsations was thought to be a case of pulseless or Takayasu's disease and investigated further.

Investigations revealed normocytic normochromic anemia with hemoglobin of 9.6 mg/dL. Erythrocyte sedimentation rate (ESR) was 86 mm at the end of 1 hour by Westergren's method and C-reactive protein (CRP) was 54.7 mg/dL (raised). Renal and liver function tests were normal. Serum thyroid-stimulating hormone was normal. Fasting lipid profile was normal. Venereal disease research laboratory test was negative. Antinuclear antibody titers by immunofluorescence assay were negative and so were anticardiolipin antibodies. Chest radiograph was normal as well. Echocardiography was normal without evidence of valvular abnormalities. Fundoscopy showed a healthy retina and disk.

Computed tomography angiography (Aortography) (Fig. 1) revealed the following:

- Circumferential thickening of the brachiocephalic trunk with near total occlusion of the right common carotid artery up to a length of 3.5 cm.
- Proximal 5 mm of right subclavian artery at its origin is opacified by contrast. Circumferential wall thickening with complete luminal wall stenosis is noted in its mid-portion (21 mm) with distal part well formed by the collaterals.
- Near-total complete occlusion of the left subclavian artery from its origin is seen with circumferential wall thickening with complete luminal wall stenosis in its proximal and middle parts.

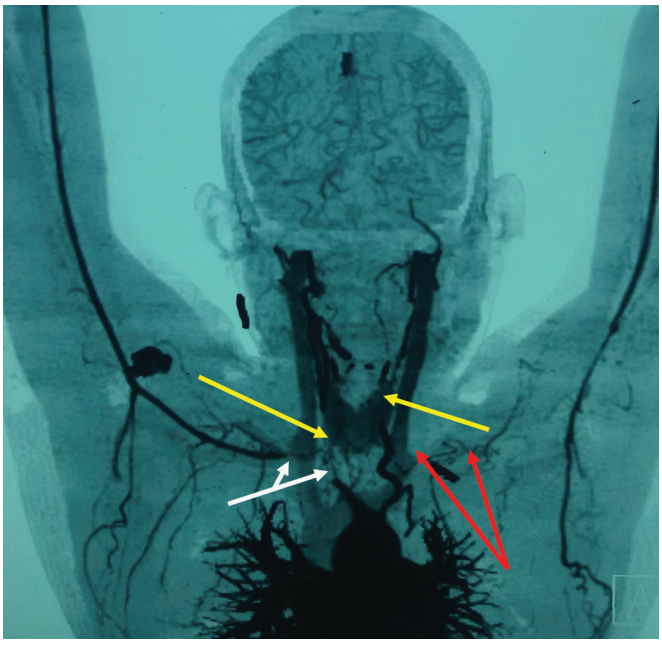

Fig. 1: Computed tomography angiography of the patient showing narrowed vessels. Yellow arrows show narrowed common carotids; white arrows show narrowed brachiocephalic trunk and consequent right subclavian artery; red arrows show narrowed left subclavian artery (when compared with the right side)

- Right vertebral artery is completely opacified.
- Renal and aorta in abdomen are normal without evidence of aneurysm.
- Circumferential wall thickening with a maximum wall thickness of 3.7 mm was noted in the distal aortic arch without significant luminal narrowing.

The arteriography confirmed our clinical diagnosis of pulseless disease as per the American College of Rheumatology (ACR) criteria. She is being treated with weight-based prednisone at a dose of 1 mg/kg. As the disease is in active phase, surgical intervention was deferred. Patient was reassessed after a month. Her acute inflammatory markers were decreased but were still raised, steroids were continued. She did not develop any new symptoms, is asked to follow-up regularly every 2 months.

The present case also raises some pertinent questions about today's clinical practices in the city. Clinical examination starts with checking of vital parameters like temperature, pulse, respiration, and BP, which appears to be missing when the patient was first hospitalized. It may be because of non-MBBS doctors working in charitable hospitals as resident doctors. Secondly, the dependency on investigations is increasing. A case of hemiplegia is first subjected to neuroimaging before clinical diagnosis, otherwise the absent pulsations cannot be missed.

DISCUSSION

Takayasu's arteritis (TA), also known as "pulseless disease," "thromboaortopathy," and "Martorell syndrome," is defined by the Chapel Hill Consensus conference on the Nomenclature of systemic vasculitis as "granulomatous inflammation of the aorta and its major branches."[1] The disease is more common in Asian population with numbers as high as 150 per million in Japan.[2] Incidence is also high in India but exact numbers are not known. An association with the tubercle bacillus was postulated as this disease was commonly seen where tuberculosis is most prevalent. However, the association is not very strong.[3] The median age of presentation is 25 years; however, approximately 25% of cases begin before 20 years of age and 10 to 20% present after 40 years of age.[4] It is characterized by chronic inflammation of arteries leading to wall thickening, fibrosis, stenosis, and thrombosis. It affects predominantly aorta and its branches. The pathogenesis of TA starts in a genetically predisposed individual with perhaps a specific hormonal milieu, followed by an exposure to unidentified antigen leading to mounting of an immunological response that targets large vessels.[3]

The patient may be completely asymptomatic at presentation and the disease may be discovered on routine check-ups when the BP is recorded or may present with devastating complications like stroke. Panja's series of 650 cases of TA, the largest series in India, reported an incidence of stroke to be 22%. Nonspecific features include fever, weight loss, arthralgia, myalgia, malaise, and anemia. As inflammation progresses, stenotic lesions develop and patient develops associated symptoms. Diminished or absent pulses, vascular bruits, hypertension, retinopathy, aortic regurgitation, congestive cardiac failure, neurological manifestation, and pulmonary artery involvement are some of the common manifestation of these patients.[5]

Infiltration with gamma delta T-cells in aortic tissues results in damage of the layers of the vessel wall by perforin. Recognition of heat shock protein 65 may result in recognition and adhesion of these cells. They have previously found restricted VαVβ gene usage of the αβT cell receptor, suggesting that a specific antigen was being targeted. More recently, restricted usage of the VγVδ genes in the infiltrating γδT cells has been reported, supporting their hypothesis, along with the expression of various costimulatory molecules necessary for T-cell activation.[6]

Some patients also had titers of antiendothelial antibodies which, in one study, was found in 18 out of 19 patients with titers 20 times greater than the normal levels.[7] Antinuclear antibodies, antineutrophilic antibodies, or antiphospholipid antibodies were all negative in all patients with TA. Tuberculosis has been particularly implicated in view of the high prevalence of infection, past or present, in affected patients,[8] largely from endemic areas. More recently, viral infection is being investigated as a trigger of vasculitis.[9]

The criteria laid down by the ACR for classification of TA has a sensitivity of 91% and specificity of 98% for the diagnosis. Three or more criteria out of six should be fulfilled for it. The criteria are:

1. Onset before 40 years of age
2. Limb claudication
3. Decreased brachial arterial pulse
4. Unequal arm BP (>10 mm Hg)
5. Subclavian or aortic root bruit
6. Arteriogram abnormality.

There is angiographic evidence of narrowing or occlusion of aorta or its primary branches, or large limb arteries.[10] Ultrasound, CT, and magnetic resonance angiography (MRA) have shown to be as good as conventional angiography for the visualization of vessels affected in TA. Magnetic resonance angiography provides high-resolution detail of vessel wall thickness and luminal calcification; also used to determine the vessel wall thickness and lumen configuration. It allows the measurement of wall enhancement as a reflection of edema and inflammation. Compared with the gold standard of angiography, 2% of stenosed vessel are overestimated as occluded in MRA.[5] Erythrocyte sedimentation rate and CRP are elevated in active disease but not in all patients, ESR being more sensitive than CRP in detecting active disease.[11] Prednisone at a dose of 0.5 to 1 mg/kg is the cornerstone of the disease in its active form. The full dose of steroids is to be taken for at least 8 to 12 weeks and followed by a gradual taper, no more than 10% of the original dose per week when remission occurs. Methotrexate, leflunomide, and azathioprine have all been used in various small-scale trials as steroid sparing agents and in patients with steroid-resistant cases, with methotrexate having an edge above the others. Small trials have shown good efficacy of anti-tumor necrosis factor agents, infliximab and etanercept, in treating patients with refractory TA.[12] Relapses occur when treatment is stopped though.

Tocilizumab is an interleukin-6 antagonist that has been found to be effective in refractory TA.[13]

Cyclophosphamide, in doses of 2 mg/kg, though toxic, is employed when other therapies fail. Evascularization procedures are done when there is:

- Critical renal artery stenosis with hypertension
- Extremity claudication limiting activities of daily living
- Cerebrovascular ischemia or critical stenoses of three or more cerebral vessels
- Moderate aortic regurgitation
- Cardiac ischemia with confirmed coronary artery involvement.[4]

Bypass surgeries have a better rate of revascularization than angioplasty and are used when the involved segment cannot be treated by angioplasty.[14,15] The mere presence of stenosis does not necessitate intervention. The gut, e.g., has such rich collaterals that even critical stenoses of the celiac, superior, or inferior mesenteric arteries usually produce no symptoms and require no surgical intervention. Moreover, many patients with arm claudication will develop collateral circulation and improve substantially over time with medical therapy alone. For upper extremity vascular insufficiency, patiently waiting for a response to medical therapy usually pays higher dividends than undertaking rapid surgical intervention. Surgical intervention should be deferred until it is in remission; procedures done during active disease often produce disappointing results.[16]

This case is reported here to increase the awareness about this pulseless disease in clinical practitioners in our country. Absent pulses and unrecordable BP should not be taken in isolation as indicators of circulatory collapse. Advanced diagnostics with modern tools and machines cannot replace the insight of the clinician. William Osler had said "The good physician treats the disease; the great physician treats the patient who has the disease."

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