

Takayasu Arteritis presenting as Hypertensive Encephalopathy

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ABSTRACT

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Hypertension with constitutional symptoms should make us suspect underlying vasculitis. We present a case of a 56 year old lady, who was diagnosed with hypertension since last 20 years, presented to us with features of hypertensive encephalopathy. Meticulous clinical examination revealed asymmetry of pulses and blood pressure between upper limbs. Further investigations helped us to make a diagnosis of Takayasu Arteritis.

Keywords: Vasculitis, Hypertensive encephalopathy, Takayasu arteritis

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INTRODUCTION

Takayasu's arteritis (TA) is a granulomatous vasculitis of the large vessels, with a predilection towards branches of aorta. Patients commonly present with headache, visual symptoms and systemic features of fever, malaise, weight loss etc.¹ Commonly observed signs are hypertension, asymmetric pulses and bruits over large vessels.² Majority of Indian patients suffer from type III TA.³ Constitutional symptoms are predominant in TA, but neurological features like headache (most common), stroke and hypertensive encephalopathy may also occur.

CASE REPORT

A 56 year old lady presented to the Emergency Department with altered sensorium of 1 day duration. She gave history of systemic hypertension for last 20 years. She had suffered an ischemic stroke at the age of 40 years with left hemiparesis and was also diagnosed to have bilateral renal artery stenosis 3 years back. She had symptoms of polyarthralgia on and off for last 20 years.

Initial evaluation showed a high blood pressure of systolic B.P 220 and non recordable diastolic pressure in the Right upper limb. Left Radial pulse was absent and Blood pressure in the Left Upper limb was unrecordable. All other peripheral pulses were present

normally. She was febrile, with no focal neurological deficits and no meningeal signs. Fundus examination showed bilateral papilloedema. Cardiovascular system examination showed Corrigan's sign on the Right, Extensive bruit were present over both carotids more on the Left. There was no renal bruit. She had an abscess over her left index finger. Clinical diagnosis of Underlying Vasculitis –Aortic Arch with occlusion of the left carotid and left subclavian, Abscess Left finger and Hypertensive encephalopathy was made.

Blood investigations showed a neutrophilic leucocytosis with a raised ESR of 62 mm 1st hour. She also had an acute kidney injury with a serum creatinine of 3.8 mg/dl. Pus culture from the wound and blood culture yielded profuse growth of Staphylococcus aureus.

Underlying vasculitis was suspected and subsequently anti ds DNA, ANCA profile, Rheumatoid factor, APLA profile were done which came out negative (**Table 1**).

Suspecting Takayasu arteritis MR Angiogram of aortic arch was done (CT angio was deferred due to Acute

Table 1. Special Tests done

Anti nuclear antibody	Negative
Anti ds DNA	Negative
C-ANCA	0.23 IU/ml (negative)
P-ANCA	0.37 IU/ml(negative)
Anti cardiolipin	Negative
TSH	1.91 uIU/ml

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Kidney injury) which showed concentric thickening of the arterial wall of the left carotid artery with segmental arterial narrowing near the origin of left subclavian artery consistent with Takayasu's Arteritis.

DIAGNOSIS AND TREATMENT

A final diagnosis of Takayasu arteritis presenting as hypertensive encephalopathy with sepsis and acute kidney injury was made. She was initiated on anti Staphylococcal antibiotics, nitroprusside and hydralazine for Blood pressure control and later calcium channel blockers. She was initiated on prednisolone at 1 mg/kg and other supportive measures for acute kidney injury. Patient improved and blood pressure became under control and renal function returned to normal.

DISCUSSION

New-onset hypertension in a patient with systemic symptoms such as fever, weight loss, and joint pain should be a clue that vasculitis may be present.⁴ Takayasu arteritis is a chronic vasculitis of unknown cause, primarily affecting the aorta and its main branches. Takayasu arteritis involves the large arteries, with hypertension developing in more than half of patients because of narrowing of the renal artery, the aorta, or its main branches and decreased elasticity. Cases are most commonly reported from Japan, China, India and other south-east Asian countries.

Neurological manifestations have been found in 52.7% of TA patients.⁵ Headache was the most common symptom (55%). The major neurological events that occurred in about one-half of the patients were TIA, cerebral infarction, hypertensive encephalopathy, paraplegia and even Moya-Moya phenomenon. These events are related to a combination of carotid and vertebral artery disease and the complication of hypertension and thromboembolism would lead to stroke. Seizure has rarely been reported as a neurological complication of TA. Seizure associated with TA has been attributed to cerebral ischemia and hypertensive encephalopathy which is occasionally associated with PRES.^{5,6}

Treatment of TA is divided into medical and surgical or endovascular therapy. Glucocorticoid therapy is the first line in medical treatment. Second-line drugs, including cyclophosphamide, Azathioprine and methotrexate, should be taken in refractory cases or in patients with recurrence of disease activity.^{7,8} Antiplate-

let or anticoagulant agents prevent thromboembolic events. Surgical or endovascular therapy of TA has been performed in critical stenosis of renal, coronary or cerebral vessels.

In conclusion, this report describes an example of how a common condition in clinical practice, hypertension, may have an unusual presentation as a hypertensive emergency with an even more unusual cause—renovascular disease with Takayasu arteritis. The correct diagnosis at the acute phase requires a thorough and open-minded evaluation of all clinical signs, symptoms, and findings, with realization that an unrecognized hypertensive emergency can lead to permanent damage.

END NOTE

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