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A RARE CASE OF TAKAYASU'S ARTERITIS PRESENTING IN CHILDHOOD

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ABSTRACT

Takayasu's arteritis is a chronic inflammatory large vessel vasculitis of unknown etiology that commonly affects adolescent female in 2nd to 4th decade of life. It usually appears to have nonspecific symptoms such as headache, fever, muscle pain, arthralgia, night sweats, weight loss, hypertension etc. Its presentation at childhood in male patient is rare. Our patient presented with irritability, convulsions, headache, vomiting, and claudication of left upper extremity but no fever. Physical examination revealed hypertension, barely palpable left radial and brachial pulsation but no abdominal bruit or carotid involvement. Plain CT brain suggestive of generalized cerebral edema. CT Angiography suggestive of narrowing of left subclavian artery & right renal artery, inflammatory thickening of renal and infrarenal part of abdominal aorta without any aneurysm. He was treated with immunosuppressive agents and shows significant improvement. Though Takayasu's arteritis is not so rare in 2nd to 4th decade of life in women but presentation in childhood particularly 1st decade is rare in male patient.

INTRODUCTION

Takayasu's arteritis, is an inflammatory large vessel vasculitis of unknown etiology that commonly affect women of child bearing age group.[1,2]The most common presenting vascular symptoms are claudication (35%), reduced or absent pulse (25%), carotid bruit (20%), hypertension (20%), carotidynia (20%), light headedness (20%) & asymmetrical blood pressure in arms (15%). Less than 10% patient may present with stroke, aortic regurgitation and visual abnormalities at the onset.[3] The most common sites of lesion in Takayasu's arteritis are aorta (65%) and the left subclavian arteries.[3] We are presenting a rare case of Takayasu's arteritis involving left subclavian artery, renal and infrarenal part of abdominal aorta and right renal artery in childhood (1st decade) in a male patient.

CASE

A 8 years male patient presented with sudden onset of vomiting, headache, irritability, seizures, claudication of left upper extremity with no history of loss of consciousness, head injury or fever. He had no past history of type 1 diabetes mellitus, hypertension, tuberculosis, heart disease or seizures. He had no family history of similar illness. His bowel and bladder habits were normal.

On examination, he was conscious but irritable. He was thinly built and malnourished. There was pallor but no icterus, cyanosis, clubbing, edema, generalized lymphadenopathy. Pulse rate was 90/min, regular at right radial artery. It was barely appreciable at left radial and brachial artery. Rest of the peripheral pulsation were well felt. No bruit heard anywhere. His blood pressure at right arm was 180/110 mm of Hg at brachial artery, left arm not recordable, right lower limb 192/120 mm of Hg and left lower limb 190/118 mm of Hg over popliteal artery. On CNS examination, higher function examination, cranial nerves, tone & power were normal. All deep reflexes brisk and planters were extensor bilaterally. Fundoscopic examination was normal. Rest of the cardiovascular system, respiratory system and per abdominal examination was normal. Laboratory evaluation revealed hemoglobin 8 gm/dl, Total leukocyte count 8600/cumm, Neutrophils 60%, lymphocytes 36%, eosinophils 2% , monocytes 2% & basophils 0%. Platelets count was 5.6 lacs/cumm. ESR was 76 mm at 1 hour, CRP was 84 mg/l. Liver function test was normal. Blood urea 22 mg/dl and creatinine 0.9 mg/dl. USG abdomen suggestive of small right kidney (4.4 length & 2.1 cm breadth) and normal left kidney (7.9 cm length, 4.4 cm breadth). Serum total cholesterol was 149 mg/dl, triglycerides 98 mg/dl. CT head revealed generalized cerebral edema. CT angiography revealed narrowing of left subclavian artery 3mm, right renal artery 1.3 mm and thickening of renal and infrarenal part of abdominal aorta. Rest pulmonary arteries, carotid arteries, brachiocehalic and right subclavian arteries were normal. No aneurysm anywhere. Echocardiography suggestive of trivial aortic regurgitation with normal left ventricular ejection fraction.

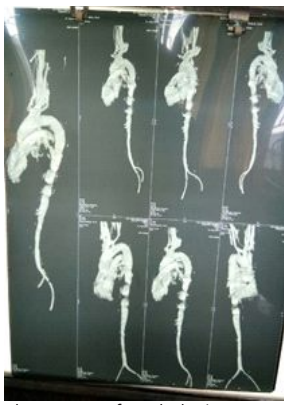

Photo 1. Left subclavian artery stenosis

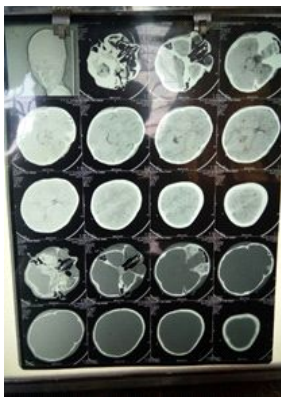

Photo 2. CT head showing cerebral edema

DISCUSSION

Takayasu's arteritis, a potentially life threatening illness is a granulomatous inflammation of large arteries. It appears have acute early phase, with nonspecific symptoms such as hypertension, headache, fever, muscle pain, arthralgia, night sweat and weight loss. Due to these nonspecific symptoms and absence of specific laboratory parameters, the disease is often unrecognized at this phase. The most common manifestations of Takayasu's arteritis are limb claudication and ischemia due to peripheral vascular involvement, hypertension from renal artery stenosis, ophthalmologic disease as manifested by retinopathy or amaurosis fugax, aortic regurgitation resulting from dilatation of ascending aorta, cardiac ischemia or congestive cardiac failure as a result of hypertensive and aortic disease, pulmonary hypertension from pulmonary arterial involvement & neurologic disease (

seizures & stroke) as a result of intra and extra-cranial arterial inflammation or thrombosis. Since large vessel biopsies are most often not possible, the diagnosis of TA is based on clinical criteria. Laboratory investigations should support the diagnosis of TA and imaging result must be confirmatory.

The presence of 3 or more of the six criteria of the American College of Rheumatology[4] is sensitive (91%) and specific (98%) for the diagnosis of Takayasus's Arteritis. Seizures, headache, hypertension and pulselessness is common presenting feature of Takayasu's Arteritis as in our case but presentation in childhood in male patient is particularaly uncommon. Involvement of intracranial vasculature is rather unusual.[5] Approximately 10%-20% of patients with Takayasu's arteritis are likely to have cerebrovascular accidents.[6] Occlusion of vertebral or carotid arteries may cause ischemic stroke. Patients with Takayasu's arteritis may also develop intracranial aneurysms.[5] Embolism of stenotic or occlusive lesions of the aortic arch and its branches, hypertension, cardiac embolism and cerebral hypoperfusion have been postulated as the mechanisms for occurrence of stroke in Takayasu's arteritis.[7] Our male patient with age of 8 years presented with headache, hypertension, seizures, claudication of left upper limb, barely palpable pulse of left upper extremity, BP not recordable at left upper arm over brachial artery, CT angiographic abnormality in the form of left subclavian artery stenosis, right renal artery stenosis with inflammatory thickening of renal and infrarenal part of abdominal aorta. He fulfilled American college of Rheumatology Criteria for Takayasu's arteritis.[4] He has type 5 disease[8] showing severe involvement of left subclavian artery, right renal artery and renal and infrarenal part of abdominal aorta. Generalized cerebral edema, seizures may be due to hypertensive encephalopathy.

Patient was treated with Prednisolone & Mycophenolate Mofetil, Aspirin and Physiotherapy. He responded well and improved symptomatically over 3-4 weeks.

So the disease should be suspected in childhood patients, though uncommon, if clinically justified. All peripheral pulses should be examined for any bruit. Immunosuppressive therapy can reduce the morbidity.

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