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Subclavian Steal Syndrome Secondary to Takayasu's Arteritis in a Young Caucasian Female

Medical Science

KEYWORDS : TAKAYASU'S ARTERITIS, SUBCLAVIAN STEAL SYNDROME

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ABSTRACT

Takayasu's arteritis, or "pulseless" disease is a rare, idiopathic, chronic granulomatous vasculitis that affects aorta and its major branches.Takayasu's arteritis mostly affects young women. The current case report focuses on an Indian rural young girl who had an attack of syncope followed by chest pain. Discrepancy in peripheral pulsations in both upper limbs and the right brachial artery BP (blood pressure) measurement not being possible, gave a hint towards the diagnosis of Takayasu's arteritis and further investigations were carried out. With Takayasu's Arteritis being a rare condition and its acute phase presentation often similar to other conditions, diagnosis is often difficult.

INTRODUCTION

Takayasu's arteritis (also known as "aortic arch syndrome", "nonspecific aortoarteritis" and the "pulseless disease") is a form of large vessel granulomatous vasculitis with massive intimal fibrosis and vascular narrowing affecting often young or middle-aged women of Asian descent. It mainly affects the aorta (the main blood vessel leaving the

Heart) and its branches. This eventually results in end organ/tissue ischemia and leads to different clinical manifestations of the disease [1, 2] The disease occurs more commonly in young females than males with peak incidence between 15 and 20 years of age. [3] It is one of the most common vasculitic disorder in India[4] and the third most common vasculitis after Henoch- Schonlein purpura and Kawasaki disease in the

paediatric age group worldwide[5-7] We present a case of Subclavian Steal Syndrome secondary to Takayasu Arteritis (TA) in a 16 year-old female Caucasian patient.

CASE REPORT

A 16 year old rural, Indian, female had a sudden episode of unconsciousness while she was doing her household chores. She became conscious spontaneously after about 3 to 5 minutes. She complained of retrosternal chest pain associated with perspiration following gain of consciousness. She was rushed to the emergency department. There was no history of headache, vomiting, fever, convulsions, numbness or weakness of any extremities, palpitations or breathlessness. Her past and family history was unremarkable.

On examination, she was conscious and well oriented. Temperature was normal, pulse-100/min and regular in left radial artery with normal force, volume and tension and condition of the arterial wall. Surprisingly no peripheral pulses were appreciated in right radial and right brachial artery. Rest all the pulsations were normal. Her blood pressure in left brachial artery was 130/80 mmHg in supine position and was not measurable in the right brachial artery. Respiratory rate was 16/min, regular. Respiratory and cardiovascular system examinations were normal.

ECG was done which showed Global ST-T changes with T inversion in precordial and limb leads. Laboratory Investigations revealed hemoglobin of 9.6 mg/dl. ESR was 98mm/h. Her electrolytes, urine analysis, renal function and liver function tests were within normal limits. Quantitative S.Troponin I was 0.593ng/ml. (Normal 0.30) and CPK-MB was 32 IU/l(Normal 0 - 24 IU/l).ANA ,c-ANCA, p-ANCA were sent on subsequent days which were negative.

Echocardiography (ECHO) was suggestive of mild Left ventricular dysfunction (LVEF 50%), RVSP 25mmHg, mild MR, mild AR, mild TR.

MSCT Ascending Aortogram was done after intravenous injection of non-ionic contrast (@ 5 ml per second) which revealed Focal Near total occlusion of 2nd part of right subclavian artery (5.8 mm length segment). Rest of SCA showed collateral reinforcement.(Image 1)

CT angiography was done which revealed occlusion of 2nd part of right subclavian artery, dilated left ventricle with thin rim of subendocardial hypodensity throughout left ventricle representing ischemic changes with normal calibre of Right coronary artery, Left anterior descending and Left circumference.(Image 2). Renal and carotid angiogram were unremarkable.

On the basis of clinical manifestations, laboratory investigations and angiogram reports a diagnosis of Takayasu's Arteritis was made.

Percutaneous Transluminal Angioplasty was planned to the Right Subclavian artery. 3.5 * 6 F guiding wire was passed, followed by balloon inflation (Cook 4 * 100 mm) at 4 ATM pressure for 10 sec. A complete 8 * 40 mm self expanded stent was inserted. Post PTA check angio revealed TIMI Grade III flow.(Image 3)

She was started on dual antiplatelets(Aspirin and clopidogrel).She was also given steroid in tapering doses. Her radial pulse was detected in right radial artery and blood pressure was 90/60 mm Hg. Within 3 months her ESR decreased to 20mm/1st hr.

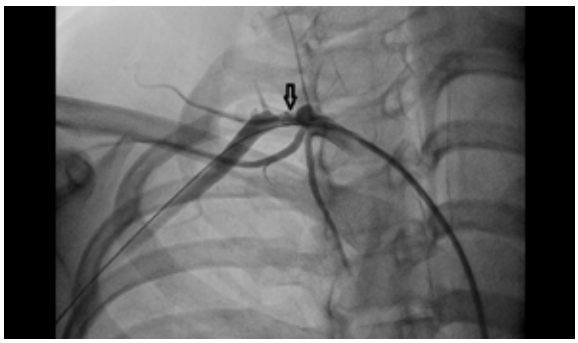

IMAGE 1- MSCT ASCENDING AORTOGRAM. ARROW INDICATING NEAR TOTAL OCCLUSION OF 2ND PART OF RIGHT SUBCLAVIAN ARTERY.

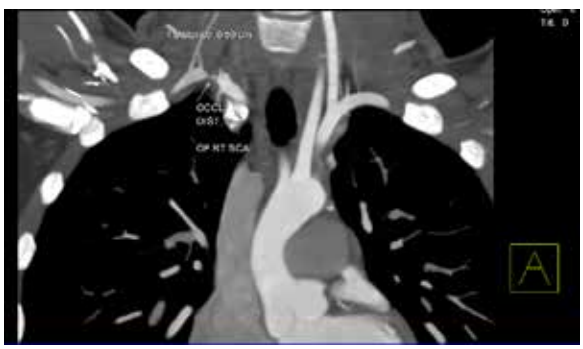

IMAGE 2- CT ANGIO DEPICTING OCCLUSION OF RT SCA.

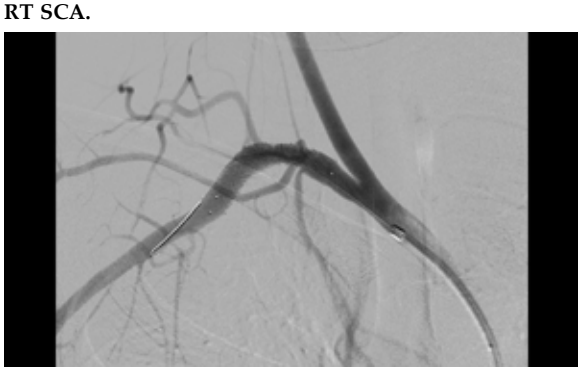

IMAGE 3- MSCT ASCENDING AORTOGRAM POST PTA TO RT SUBCLAVIAN ARTERY.

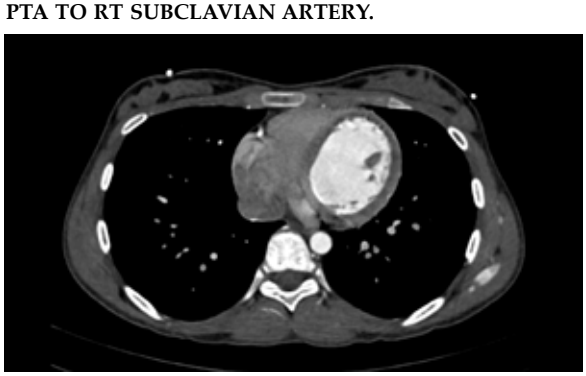

IMAGE 4- CARDIAC CT IMAGING SHOWING DILATED LV WITH THIN RIM OF SUBENDOCARDIAL HYPODENSITY.

DISCUSSION

Thus with the clinical manifestations and their radiological co-relation a diagnosis of Takayasu arteritis can safely be established in the above case. The first case of Takayasu's arteritis was described in 1908 by a Japanese ophthalmologist, Mikito

Takayasu at the annual meeting of the Japan Ophthalmology Society [6, 7]. Takayasu described a peculiar "wreath-like" appearance of the blood vessels in the back of the eye (retina). The Worldwide incidence of Takayasu arteritis is estimated at 2.6 cases per million per year. Although the disease has a worldwide distribution, it is observed more frequently in Asian countries such as Japan, Korea, China, India.

The erythrocyte sedimentation rate (ESR) probably remains the most reliable marker of disease activity, but even this test is not helpful in a sizeable number of patients who have active arterial inflammation but normal ESR. The Ishikawa criteria [8] (1986) have been useful in defining Takayasu arteritis. The above case satisfies the two major criterias with age less than 40 years at the onset of symptoms or diagnosis and involving lesions in left and right subclavian artery as determined by aortography. The minor criterias consist of annuloaortic ectasia or aortic regurgutation as shown by angiography or ECHO and pulmonary artery,left mid common carotid, distal brachiocephalic trunk, descending aorta or abdominal aorta lesions. However this criterion was more useful in diagnosing the disease in Japanese population.

The American College of Rheumatology [9] has given their criteria in 1990 for diagnosing the disease (3/6 are necessary): age 40 or <40years at disease onset, claudication of extremities, decreased pulsation of 1 or both brachial arteries, difference of 10 mmHg in systolic BP between both arms, bruit over 1 or both subclavian or abdominal aorta and angiographic criteria must show narrowing or occlusion of the entire aorta, its primary branches or large arteries in the proximal upper or lower extremities, these changes are not due to arteriosclerosis, fibromuscular dysplasia or similar causes and the changes are focal or segmental. The ACR criterion gives a greater flexibility to account for the variability in actual clinical practice.

In the setting of severe proximal subclavian artery stenosis or occlusion, typically due to atherosclerotic disease, insufficient flow may be present to sustain the ipsilateral arm. In this case, the branches of the subclavian artery may be recruited to provide collateral retrograde flow to the upper limb. Of greatest relevance for present purposes is the confluence of the vertebral arteries at the basilar artery and its subsequent communication with the circle of Willis, which allows the ipsilateral vertebral artery to provide flow in a retrograde manner from the contralateral vertebral artery or from the anterior cerebral circulation. With exercise, innate and metabolite-induced vasodilatation leads to a drop in peripheral resistance in upper-limb vessels, and the mismatch between arterial inflow and metabolic demand may lead to claudication of the arm. In these patients, neurologic symptoms consistent with cerebral or brainstem ischemia may develop. (Dizziness, vertigo, blurred vision, diplopia, and near-syncope) upon exercise of the ipsilateral arm. This girl developed syncope while she was doing her household chores. As she had narrowing of her subclavian artery proximal to right vertebral artery, the cause of her syncope may be subclavian steal syndrome secondary to Takayasu arteritis.

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CONCLUSION

Takayasu's arteritis is a rare systemic vasculopathy that can progress to vital organ ischemia. The aim of treatment must be the control of disease activity and

the preservation of vascular competence, with minimal long term side effects. Patients with disease that carries a good prognosis should not be put at risk by treatment that is more harmful than the disease itself. Current evidence favours the use of steroids and

immunosuppressive drugs. As the treatment for Takayasu's arteritis may be associated with substantial side-effects, more accurate means of gauging disease activity are required.The present case underscores that despite the very low incidence of TA in Caucasians (0.8/1,000,000), this large-vessel vasculitis of unknown etiology should always be considered in the differential diagnosis of subclavian steal syndrome in Caucasian women aged less than 40 years.

REFERENCES

1) Harrison's principles of internal medicine.
2) Braunwald's textbook of cardiology.
3) Saab, Fadi;giugliano, Robert P; Giugliano, Gregory R. 2009-01-01.
4) VR Joshi, Gayatri Mittal. Vasculitis-Indian perspective. JAPI june 2006:vol.54
5) Jain S, Kumari S, Ganguly NK, Sharma BK. Current status of Takayasu arteritis in India. Int J Cardiol. 1996; 54 (Suppl.):S111–6.
6) Murangan MN, Bavdekar SB, Desmukh H, Tripathy M, Vaswani R. Study of Takayasu's arteritis in children: Clinical profile and management. J Post Grad Med. 2000; 46: 3–8.
7) Brogan PA, Dillon MJ. Vasculitis from the pediatric perspective. Current Rheumatology Rep. 2000; 2: 411–6.
8) Ishikawa K; Natural history and classification of occlusive thromboaortopathy (Takayasu's disease).JACC Circulation. 57 1978:27-35.
9) American College of Rheumatology.