Takayasu’s Arteritis – Stroke as an Initial Presentation

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Abstract

A 22 year old lady presented with history of sudden onset of inability to move her left upper and lower limbs with deviation of the angle of the mouth to the right side. No previous history of fever, joint pain, or other systemic manifestations were present. Magnetic resonance angiography revealed long segment narrowing of common carotids, complete occlusion of the first and second portions of the left subclavian, right brachiocephalic, and proximal 4 cms of vertebral artery. However, the thoracic and abdominal aorta along with renal arteries were normal. The patient improved with oral steroids and anticoagulants.

Introduction

Takayasu’s arteritis is a chronic inflammatory disease of unknown cause characterised by granulomatous vasculitis of medium and large arteries, principally the aorta and its branches1. Takayasu’s arteritis classically exhibits a triphasic pattern of expression consisting of systemic non-vascular phase; a vascular inflammatory phase, and a quiescent “burnt out” phase. However, this classic presentation holds true only in a minority of patients. We report a case in which the complication, i.e, stroke, heralded the onset of the disease.

Case report

A 22 year old lady presented to us with history of sudden inability to move her left upper and lower limbs, deviation of the angle of mouth to the right side associated with giddiness. She gave history of few transient episodes of loss of consciousness with spontaneous recovery over the past 2-3 months. There was no past history of fever, joint pain, or other systemic manifestations.

On examination, the patient was pale, moderately built, and nourished. She was afebrile. Her left radial and brachial pulses were absent and were feeble on the right side. Blood pressure in right upper limb was 118/70 mm Hg and left upper limb 102/64 mm Hg. Blood pressure recorded in the lower limbs – 140/80 on both the sides. There was a thrill associated with a bruit over both the carotid arteries. All the lower limb pulses were felt. She had a left upper motor neuron facial palsy with a left dense hemiplegia (power grade 0/5) Cardiovascular examination showed a grade II ejection systolic murmur over aortic area. There was no renal vascular bruits. Optic fundi did not reveal any abnormality.

Her ESR was 110 mm/hr, total counts 15,500, C-reactive protein 18 microgram/ml. Chest X-ray was normal and ECG was within normal limits. Anti nuclear antibody (Immunoflorescence), lupus anticoagulant, and anticardiolipin antibodies were absent. Echocardiography revealed normal LV function. Carotid doppler showed 70% stenosis of proximal right common carotid artery and 60% of whole of left common carotid artery. Right vertebral artery was dilated resembling the common carotid. Magnetic resonance imaging of brain showed infarction of genu of right internal capsule, right lentiform nucleus, and corona radiata (Fig 1 and 2). Magnetic resonance angiography revealed long segment narrowing of common carotids, complete occlusion of first and second portions of left subclavian, right brachiocephalic, and proximal 4 cms of left vertebral artery. However, thoracic, abdominal aorta and renal arteries were normal (Fig. 3).

The patient was treated with steroids and anticoagulation. In 6 months of follow-up, the patient has been doing well with return in limb power to grade 4/5. No evidence of hypertension or other systemic manifestations have been so far detected.

Discussion

Takayasu’s arteritis also known as “pulseless disease”, thromboaoortopathy and “Martorell syndrome” is characterised by chronic vessel inflammation leading to wall thickening, fibrosis, stenosis, and thrombosis. It affects predominantly aorta and its branches.
Symptoms depend on end organ ischaemia. Published descriptions of this arteritis date back as far as 1830. In 1905, Takayasu, Professor of Ophthalmology, presented the case of a 21-year-old woman with characteristic fundal arterio-venous anastomosis. Takayasu’s arteritis is rare, but commonly seen in Japan, South East Asia, India, and Mexico. It has been included in the list of intractable diseases by the Japanese Government and to date 5,000 patients have been registered.

Takayasu’s arteritis is characterised by two-stage disease “pre-pulseless” with non-specific inflammatory features followed by a chronic phase characterised by vascular insufficiency. The disease presents in the 2nd or 3rd decade of life. A study in India gave a mean age of symptom onset of 24 years and age at diagnosis of 28.3 years. Non-specific features include fever, weight loss, arthralgias, myalgia, malaise, and anaemia. As inflammation progresses, stenotic lesions develop and patients develop associated symptoms. Diminished or absent pulses, vascular bruits, hypertension, retinopathy, aortic regurgitation, congestive cardiac failure, neurological manifestations, and pulmonary artery involvement are some of the common manifestations of these patients. Panja’s series of 650 cases of Takayasu’s arteritis, the largest series in India, reported an incidence of stroke as 22%.

1990 American College of Rheumatology criteria for diagnosis of Takayasus Arteritis:

1. Age of onset < 40 years
2. Claudication of extremities.
3. Decreased brachial artery pressure.
4. Blood pressure difference >10 mm Hg
5. Bruit over subclavian arteries and aorta.
6. Aortogram abnormalities.

At least 3 of the above 6 criteria are to be met for the diagnosis.

Tuberculosis has been implicated in the aetiology and also as an important differential, in view of the high prevalence of infection, past or present, in affected patients. A viral
trigger to the vascular inflammation has also been postulated. The association with certain HLA alleles in various populations has strengthened the argument for an autoimmune process, though no specific autoantigens have been identified. Serum concentration of IL-1, IL-6, and RANTES are elevated during disease activity and may contribute to vasculitis.

Ultrasound, CT, and recently magnetic resonance angiography (MRA) has shown promise in the diagnosis of Takayasu’s arteritis. MRA provides high resolution detail of vessel wall thickness and lumen configuration. It allows the measurement of wall enhancement as a reflection of oedema and inflammation. Compared to the gold standard of angiography, 2% of stenosed vessels are over estimated as occluded on MRA. By reduction of enhancement on follow-up MRA also serves as a surrogate marker for disease activity.

Steroids are the mainstay of treatment for Takayasu’s arteritis. Approximately half of the patients respond to steroids. Steroid unresponsive patients can be treated with cytotoxic drugs including cyclophosphamide, azathioprine, and methotrexate. Treatment should aim to control disease activity, preserve vascular competence with minimal long term side effects. Surgical treatment is offered to those with severe stenosis of renal artery, extremity claudication, stenosis of 3 or more cerebral vessels, or evidence of coronary artery involvement. Cumulative survival at 5 years after disease onset was 91%, and after 10 years the figure was 84%.

Our patient had no past history of systemic manifestations like fever, joint pains, and weight loss. Neurological deficit heralded the onset of disease. Patient showed good response to steroids and anticoagulants, with return of limb power to grade 4/5. Also the role of magnetic resonance angiogram needs to be highlighted. It is a non-invasive procedure, provides high resolution detail of vessel wall thickness and lumen configuration. In our patient, MRA revealed no involvement of thoracic or abdominal aorta and renal arteries – an uncommon presentation.

References