

Takayasu`S Arteritis Presenting as Stroke: Importance of Bedside Examination in Diagnosing Rare Cases



Medical Science

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ABSTRACT

Takayasu's arteritis (TA) is a rare chronic granulomatous vasculitis of the aorta and its branches and is associated with considerable morbidity and premature mortality. Initial symptoms and signs are non-specific. High index of suspicion is needed to make the correct diagnosis. Only 10% of cases present as CVE. We are presenting the case of 34 year female presenting as Hemiplegia on left side and having total occlusion of common carotid on right side. This case rare and it can be easily diagnose by bedside examination and delay due to investigation can be avoided especially in periphery.

Introduction

Takayasu's Arteritis is a chronic, idiopathic, inflammatory disease that primarily affects large blood vessels such as aorta and its branches.^{1,9} Neurological symptoms occur in about 50% of cases but stroke occurs in about 10% of patients with Takayasu's Arteritis^{2,8}. We report this case because it is rare and important cause of hemiplegia in young patients. We can easily diagnose this entity by proper bedside examination and delay in treatment in periphery where investigations are not so easily available can be avoided. Only palpating all peripheral pulses and taking blood pressure in all four limbs can diagnose takayasu arteritis.

Case Report

A 34 year old female presented to us with the history of sudden inability to move her left upper and lower limbs, deviation of the angle of mouth to the right side, associated with headache. Episodes of focal convulsions also present. There was no past history of fever, joint pain, other systemic manifestations.

On examination, the patient was thin built, afebrile, her right carotid, right brachial, and radial, right femoral, popliteal, dorsalis pedis pulses were absent. Left carotid was feeble with thrill present. Blood pressure in right upper limb was unrecordable and left UL 100/60mm/Hg. Blood pressure recorded in lower limbs -128/85 on left side and not recordable on right side. There was a thrill associated with a bruits over the left carotid artery. She had a left upper motor neuron facial palsy with a left dense hemiplegia (power grade 0/5). Cardiovascular examination showed normal findings, there was no renal vascular bruits, optic fundi did not reveal any abnormality. Her ESR was 40 mm/hour, total counts 12,500 cells/ μ l, chest X ray was normal and ECG was within normal limit. CT brain revealed acute infarct in right MCA territory. Carotid doppler revealed right common carotid artery stenosis 100% and mild atherosclerotic changes in left common carotid. However thoracic, abdominal aorta and renal arteries were normal. Patient was managed with steroids, antiplatelets, lipid lowering agents and anti convulsants. Patient was referred to higher center as per relatives wish.

Discussion

Takayasu's Arteritis is also called as pulseless disease, is a chronic, idiopathic, inflammatory disease that primarily affects large blood vessels like aorta and its branches¹. Takayasu, a Japanese ophthalmologist, first described the clinical entity in 1908 in a young female with retinal changes³. Latter Shimzu and Sano, in 1928, described it as pulseless disease³. Its estimated incidence is 2.6/100,000/year¹. The disease has worldwide distribution but is more common in Japan, India and China². It affects predominantly females, with a male to female ratio of

1:9 but the difference is much less pronounced in India, where the male to female ratio is 1:1.5-32. Pathogenesis of the disease remains elusive but there is an indirect evidence of an autoimmune process. Some association with sarcoidosis and inflammatory bowel disease has also been noted^{2,4}. Certain HLA antigens, like HLA-Bw 5 and B 21, are also more common in patients with Takayasu's Arteritis as compared to the general population². Takayasu's Arteritis has a variable presentation. About 15-50% of the patients have nonspecific symptoms like fever, malaise, myalgia, weight loss and arthralgia². One third of the patients are anemic and 10-30% have various cardiac symptoms². It is the most common cause of renovascular hypertension in India². About half of the patients have neurologic symptoms, the most common being visual symptoms². Strokes occur in 10% of the patients^{2,8,9}. The most common presentation is hemiparesis and hemihypesthesia while the commonest sites of involvement are the basal ganglia and watershed zones⁵.

1990 American college of Rheumatology criteria for diagnosis of TA;⁸

- 1) Age of onset < 40 years
- 2) Claudication of extremities
- 3) Decreased brachial artery pressure
- 4) B.P differences > 10 mm/Hg
- 5) Bruits 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 population has strengthened the argument for an autoimmune process. Ultrasound, CT and MRA have shown promise in the diagnosis of TA. Steroid are the main stay of treatment for TA, approximately half of the patients respond to steroids. Steroid unresponsive patient, can be treated with cytotoxic drugs including cyclophosphamide, azothioprine and methotrexate and mycophenolate mofetil⁸. 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 cerebral vessels or evidence of coronary artery involvement. Cumulative survival of 5 years after disease onset was 91 % and after 10 years the figure was 84%.

Conclusion

This case is presented because TA presenting first time as hemiplegia in only 10% of patients. It is important but rare cause of CVE in young females. This case is also presented mainly to em-

phasise the importance of looking for peripheral pulses and recording blood pressure in all four limbs at least in young stroke patients. Takayasu's Arteritis should be considered as a possibility in all young patients (especially females) who present with stroke as this is fairly responsive to medical management. This is necessary to prevent any further systemic complications, which may be devastating.

References

1. Gail SK, Claire WH, Joseph G, et al. Takayasu's Arteritis. *Ann. Intern. Med.*, 1994;120:919-29.
2. Jain S, Kuman S, Gaunguty NK, et al. Current status of Takayasu's Arteritis in India. *Int. J. Cardiol.*, 1996;54: S111-16.
3. Sharma BK, Jam s, Sun S, et al. Diagnostic criteria for Takayasu's Arteritis. *Int. J. Cardiol.*, 1996; 54: S141-47.
4. Hunder G. Vasculitis: Diagnosis and therapy. *Am. J. Med.*, 1996, 100: S37-45.
5. Naritomi H. Takayasu's Arteritis. In: Bogousslasky, J, Caplan, L, eds, *Stroke syndromes*. 1st ed. New York, Cambridge University Press, 1995; pp. 437-42.
6. Love BB, Orenca AJ, Biller J. Stroke in children and young adults: overview, risk factors and prognosis. In: Bitter J, Mathews KD, Love BB, eds, *Stroke in children and young adults*. 1st ed. Boston, Butterworths- Heinemann, 1994, pp.1-13.
7. Arend WP, Michiel BA, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu's Arteritis. *Arthritis Rheum.* 1990;33:1 129.
8. KELLYS TEXTBOOK OF RHEUMATOLOGY 9th edition page no 1473
9. HARRISONS Textbook of medicine 18th edition page no2796