

A Rare Case Report of A Pulseless Male Patient With Sequale of Takayasu's Arteritis.

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Abstract: Cardiogenic embolism is a major cause of stroke and often leads to significant morbidity and mortality. Despite the recent advances in our understanding of the pathophysiology of stroke and its risk factors, diagnosis and therapy; some case scenarios still present a real challenge for the treating physicians. We report a case of a 38 year old male patient presenting with multi-territory cerebral infarctions due to a left ventricular mobile thrombus complicated by pulseless right arm later investigated and diagnosed to be takayasu's arteritis. Gradual introduction of anticoagulation and steroids coupled with a multidisciplinary team approach advocating careful daily clinical assessment of the patient and regular echocardiographic and neuroimaging studies have resulted in a better management and achievement of therapeutic goals.

Keywords: Stroke, Cardioembolism, Mobile thrombus, Takayasu's arteritis, Echocardiography, Multidisciplinary approach

I. Introduction

Embolic stroke is a leading cause of morbidity and mortality. The heart is an important source for the development of embolic strokes. About 15-20% of all ischemic strokes are cardioembolic. Establishing the heart as a source for a patient's embolic stroke carries important prognostic and therapeutic implications. Patients with such types of stroke often suffer significant morbidity and mortality and have a high risk of recurrence. The presence of takayasu's arteritis leading to coronary artery disease followed which a left ventricular thrombus, which carries additional risks including the risk of carotid atherosclerotic plaque. Managing patients of takayasu's arteritis along with left ventricular thrombi causing an embolic stroke as we present in this report.

II. Case Report

A 38-year old right-handed male with a history of smoking and alcohol for 20 years, presented with severe headache which was localised mainly to temporal region associated with vertigo, followed by blurring of vision since 9 days and slurring of speech along with right sided weakness since 5 days. He had no history of chest pain or palpitations. He did, however, give a history of a recent visit to another medical practitioner for the same but did not get any relief. Details of his management during that visit were not available. Examination showed blood pressure on left hand in supine position of 140/90 mmHg, regular heart rate of 90 per minute, and no peripheral pulse on right hand, &clubbing grade 3, bruit over carotid and subclavian artery and Glasgow coma scale (GCS) of 15, expressive aphasia, with right sided hemiparesis and left homonymous hemianopsia. Cardiac examination as well as ECG, cardiac enzymes and basic laboratory investigations was unremarkable. Initial CT brain showed a occipital infarct and fronto temporal infarct on left side. B/I carotid colour Dopplershowed soft plaque in cca & ica on right side intima media thickened in the arteries of the neck and decreased velocity in cca, ica& eca. The patient was admitted to the high dependency unit of the general medical ward with regular assessment of his GCS.

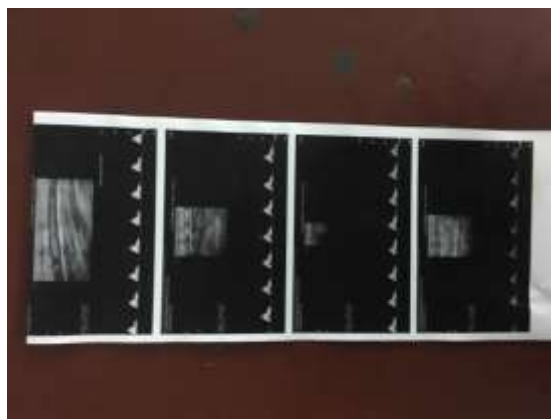
The patient's initial CT brain showed multiple areas of ischemic infarcts, the largest of which is in the distribution of the left middle & posterior cerebral artery.

Echocardiography revealed poor overall systolic function (ejection fraction 20%) with global hypokinesia, severe lv dysfunction and a mobile thrombus measuring about 2.6 cm × 1.4cm in the apical region of the left ventricle. The patient was started on 100 ml of Mannitol 20% three times daily with daily monitoring of the electrolytes profile in addition to regular physiotherapy. Forty-eight hours after the onset, he was started on weight-adjusted low molecular weight heparin (LMWH) at a prophylactic dose (enoxaparin 40 mg/day) and clopidogrel 75 mg once a day. He had minimal improvement in his neurological status. His thrombophilia workup (including tests for protein S, protein C and anti-thrombin levels and activities sent before the initiation

of LMWH, as well as molecular testing for factor V Leiden and prothrombin gene mutations) were unremarkable.



Doppler Report



Echo showing the LV thrombus.

Patient was further evaluated for the pulseless right arm by doing Colour Doppler of the right arm in order to rule out takayasu's arteritis, which showed reduced blood flow velocity, low resistance and vessel wall thickening.

Daily assessment of the patient by a multi-disciplinary team including a neurologist, a cardiologist and a hematologist in addition to the primary internist was adopted. A weekly echocardiogram was used to assess the thrombus size while weekly neuro-imaging with CT scans was initially used to assess the infarcted areas, especially with regards to hemorrhagic transformation. With no progression in the small areas of hemorrhagic transformation on CT scans, the dose of enoxaparin was upgraded to 40 mg twice daily and then after another week, it was increased to 60 mg twice daily with weekly monitoring of anti-Xa levels. Warfarin was introduced with a target INR of 2.0 to 3.0. Follow up echocardiographic studies demonstrated a significant regression in the thrombus size without any evidence of further systemic embolization. The patient was discharged after 7 weeks of hospitalization with NIH Stroke Scale score of 15 and a modified Rankin Scale score of 5.

III. Discussion

Cardiogenic embolism is recognized to account for one in six strokes^{1,2}. Common causes of cardiac embolization include atrial fibrillation, ischemic heart disease, rheumatic heart disease and prosthetic cardiac valves^{3,4,5,6}. Left ventricular thrombi (a frequent complication of transmural acute myocardial infarction, likely to be the case in our patient)^{7,8}, are second to atrial thrombi from atrial fibrillation as a cause of cardiogenic embolization. These are easily detectable by two-dimensional echocardiography, although false-positive studies exist⁹. Protruding configuration and free mobility of the thrombus and the presence of adjacent hypokinesia indicate a high embolization potential^{10,11}. The presence of a potential embolic source does not, by itself, justify

the diagnosis of cardiogenic embolism in this patient. However, the abrupt onset of maximal deficit, the presence of multiple brain infarcts involving different vascular territories along with the presence of a large left ventricular thrombus indicate that the cause of the infarcts in this patient are indeed cardioembolic. Takayasu's disease is histologically characterized by irregular thickening of the aortic wall and its branches. Early lesions consist of a mononuclear infiltrate of the vascular adventitial vasa vasorum. Later, there may be marked mononuclear infiltrate of the arterial media, accompanied in some cases by granulomatous lesions filled with giant cells and patchy media necrosis. With disease progression even after treatment with steroids, inflammatory reaction is predominantly characterized by collagen fibrosis affecting all layers of the vessel wall (especially the intima) accompanied by lymphocytic infiltration. This is the mechanism by which constrictions of the coronary arteries may lead to AMI¹².

1990 Criteria of American College of Rheumatology for the Classification of Takayasu Arteritis

Criteria	Definition
Age at disease onset in year	Development of symptoms or findings related to Takayasu arteritis at age <40 years.
Claudication of extremities	Development and worsening of fatigue and discomfort in muscles of one or more extremity while in use, especially the upper extremities.
Decreased brachial artery pulse	Decreased pulsation of one or both brachial arteries
BP difference >10mmHg	Difference of >10mmHg in systolic blood pressure between arms
Bruit over subclavian arteries or aorta	Bruit audible on auscultation over one or both subclavian arteries or abdominal aorta
Arteriogram abnormality	Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due arteriosclerosis, fibro-muscular dysplasia, or similar causes: changes usually focal or segmental

The cause of coronary artery disease in a young patient with a pulseless right arm was further evaluated and was to our surprise a rare entity Takayasu's arteritis. Being such a rare presentation came to us a challenge, which further evaluated with the help of imaging techniques.

Upon discharge our patient was treated with warfarin 2 mg/day (to keep INR in a range of 2-3), aspirin 100 mg/day, methylprednisolone 40 mg/day, clopidogrel 75 mg/day, amlodipine 10 mg every 12 hours, nebivolol 10 mg/day, hydrochlorothiazide 25 mg/day and pantoprazole 40 mg/day and the association with cyclosporine was recommended.

Conclusion

The clinical presentation of patients with Takayasu's arteritis can be as varied as the location of the affected arteries. The cases presented with AMI described in the literature are rare; in the case of our patient it was an Anterior AMI. Left ventricular thrombus complicating ischemic heart disease and causing systemic embolism is a well-recognized clinical phenomenon which requires anticoagulation. The treatments should be focused not only to prevent progression of the disease, but also to improve the quality of life. From the beginning treatment has been based on corticosteroids, oral anticoagulants, which may be associated with immunosuppressants such as cyclosporin. Early diagnosis and optimal treatment play a key role in the patient's outcome and disease progression.

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