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Acute Psychosis and Takayasu Vasculitis

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ABSTRACT

Takayasu arteritis is one of the large vessel vasculitis with variable clinical features in Indian patients. It is common in males and may be associated with tuberculosis and often presents with large vessel wall inflammation, obstruction and aneurysm. A 23-year-old female presented with features of acute psychosis and during evaluation and following electroconvulsive therapy she was found to be tachycardic and pulse less in one arm. Further CT angiogram proved features of aortoarteritis with raised inflammatory markers and her infective screen including TB came back as negative. She was treated with steroids, losartan and corticosteroids and Methotrexate was added as steroid sparing immune suppression. She remains disease free and doing well. Literature review suggests increasing incidence of this vasculitis and early effective treatment helps to reduce morbidity and prolong disease free remission.

Keywords: Takayasu vasculitis, Occluded carotid, Depression

INTRODUCTION

Takayasu arteritis is the commonest type of large vessel vasculitis. Ishikawa defined clinical groups based on the natural history and complications of the disease [1]. The four most important complications were defined as Takayasu retinopathy, Secondary hypertension, aortic regurgitation, and Aneurysm formation. (Each graded as mild/moderate or severe at diagnosis). Different ethnic populations manifest with variable disease presentation as illustrated by Moriwaki, et al. in their study of Indian and Japanese patients [2]. The Japanese patients were mostly female presenting with fatigue, dizziness, vertigo, pulselessness, more aortic regurgitation, reflecting involvement of the aortic arch and its main branches. This was very different compared with the Indian patients [2]. They tended to present with headache and hypertension due to vasculitis affecting the abdominal aorta and renal vessels.

CASE PRESENTATION

A 23-year-old female presented with two weeks history of talking to self, agitated and insomnia and found wandering on the streets. There was no history of fever, headache, or trauma. She was in acute psychosis. She was married and was in a difficult relationship. She was treated with oral antipsychotics. Her basic blood investigations were normal except ESR of 38 mm/hr. As she did not respond to the treatment she was given electroconvulsive therapy (ECT) and following ECT she was found to have tachycardia which settled afterwards. ECG following ECT showed T inversion in inferior leads.

Following cardiology review, she was found to have carotid and subclavian bruit and her CT angiogram showed occluded left common carotid and left subclavian artery, stenosed celiac axis bulb and diffusely thickened aorta. Echo showed good left ventricular function. Her immunology all came back normal. Quantiferon test for TB was

negative. Her chest imaging was normal. The diagnosis was Large vessel vasculitis-Takayasu arteritis pattern with schizophrenia. She was commenced on immunosuppression.

This is a unique presentation and although it was chance association, asymptomatic vasculitis presenting as an acute psychosis is unexplained.

CLINICAL FEATURES

The disease commonly presents in late 20s often with a long delay in diagnosis. The National Institute of Health study by Kerr, et al. suggested that the delay in diagnosis was longer in juveniles, being up to four times that of adult patients [3]. Hypertension was the commonest mode of presentation (51.3%) and was detected in 82 patients (77.4%) at the time of presentation in a study by Jain, et al. Systemic features include fever, night sweats, malaise, weight loss, arthralgia, and anemia [4]. As the inflammation progresses and vessel gets occluded, more characteristic features appear with the development of collaterals.

The most commonly affected vessels are the subclavian and common carotid arteries. Stenotic lesions are found in >90% of patients, whereas aneurysms are reported in approximately 25%. Pulmonary arteries are involved in up to 50% of patients and it is important to look specifically for evidence of aortic valve regurgitation and coronary arteritis [5]. In Indian patients, vasculitis generally occurs in the abdominal aorta affecting renal arteries and may extend into the thoracic aorta within a decade [2].

Takayasu arteritis can be divided into the following six types based on angiographic involvement based on Angiography [5]:

- **Type I** - Branches of the aortic arch
- **Type IIa** - Ascending aorta, aortic arch, and its branches
- **Type IIb** - Type IIa region plus thoracic descending aorta
- **Type III -** Thoracic descending aorta, abdominal aorta, renal arteries, or a combination
- **Type IV** - Abdominal aorta, renal arteries, or both
- **Type V** - Entire aorta and its branches

Etiology/Demographics

Although the definite etiology is unknown, the major pathology is inflammatory, with several proposed factors including spirochetes, mycobacterium tuberculosis, and streptococcal organisms and circulating antibodies due to an autoimmune process. Although Mycobacterium tuberculosis has been implicated in the pathogenesis of Takayasu's arteritis (TA), there is no direct evidence substantiating the association [6].

Takayasu arteritis is observed more frequently in patients of East Asian or Indian descent. Approximately 80% of patients with Takayasu arteritis are women [7]. Takayasu arteritis is progressive or relapsing/remitting and requires immunosuppressive treatment for disease remission and maintenance.

Prognosis

The complications of Takayasu arteritis can be severe and although medical and cardiothoracic interventions have improved, still lot more to be done to improve the prognosis. Morbidity is substantial with 74% of patients reporting considerably compromised daily activities and 23% of patients unable to work Mortality rates vary based on different approaches to the treatment. In India, 10-year survival is reported to be 80% - 85% [8,9]. Patients can develop rapidly expanding aneurysms, suffer from pulmonary hypertension or aortic rupture. Cardiac complications are common, and aortic valve insufficiency, cardiac ischemia and myocardial infarction complicated by cardiac failure are frequent causes of death [10].

Reasons for poor outcome

The relatively poor outcome for patients with this disease is due to lack of awareness, heterogeneous presentation, and rarity of the disease itself. An inadequate approach to ischemic symptoms in children and young adults, and a failure to recognize the importance of diminished pulsation in the limbs during examination are also important contributing factors.

Differential diagnosis

The differential diagnoses include other causes of large vessel vasculitis:

1) Inflammatory aortitis (tuberculosis, rheumatoid arthritis, spondyloarthropathies, Kawasaki disease, and giant cell arteritis);
2) Developmental abnormalities (coarctation of the aorta and Marfan syndrome).

Most of these have specific features for diagnosis, but tuberculosis has remained an enigma. The incidence of rupture and bleeding complications of aneurysmal Takayasu arteritis is low. Syphilis tends to affect older age group, affecting the arch and ascending aorta. Hypertension due to fibromuscular dysplasia is to be kept as differentials.

Treatment

The aim of treatment must be the control of disease activity and the preservation of vascular competence, with minimal long-term side effects. Currently, as the best evidence base suggests, corticosteroids are the standard form of treatment, to which 50% respond, and Methotrexate is used as a steroid sparing drug. Only 25% of patients with active disease may not respond to current treatments and care should be taken to avoid prolonged immunosuppression in the absence of clinical benefit. Mycophenolate has also been used to control the inflammatory disease with good results. Most patients will receive immunosuppression and thereby achieve effective disease control and reduce the burden of surgery and complications.

Infliximab and Tocilizumab are cytokine inhibitors found to be useful in managing the activity of Takayasu. In the retrospective multicenter study of outcome of 49 TA patients, treated by TNF-α antagonists (80%) or tocilizumab (20%) and fulfilling ACR criteria, the overall response (i.e., complete and partial) to biological-targeted treatments at 6 and 12 months were of 75% and 83%, respectively. The 3-year relapse free survival was of 90.9% over biologic treatment period compared to 58.7% with DMARDs [10,11].

Surgical treatment

Indications for surgery include hypertension with critical renal artery stenosis, extremity claudication, cerebrovascular ischemia or critical stenosis of three or more cerebral vessels, moderate aortic regurgitation, and cardiac ischemia with confirmed coronary artery involvement. In general, surgery is recommended at a time of quiescent disease to avoid complications, which include restenosis, anastamotic failure, hemorrhage, and infection.

Follow up

Takayasu arteritis is a systemic vasculopathy that can progress to cause vital organ ischemia. Hence, long-term follow-up is recommended. Magnetic resonance angiography (MRA) is now being used in the evaluation of large vessel vasculitides [12,13], It provides high resolution images of vessel wall thickness and lumen configuration, and allows the measurement of wall enhancement as a reflection of active inflammation. The reduction of enhancement on follow up is presumed to reflect reduced inflammatory activity. Therefore, MRA is likely to be used increasingly as an accurate follow up tool [14]. There is still lot of uncertainty with regard to the disease onset and course, and one fourth of patients may end up having progressive disease.

Outcome and implications for the case presented

This girl has now been diagnosed prior to any organ specific complications and on regular treatment with follow-up. When she plans to get pregnant, there will be some implications regarding methotrexate. Obviously, the disease will have to be controlled with safer medications.

DISCUSSION AND CONCLUSION

Although asymptomatic presentation of Takayasu vasculitis in not uncommon, presenting with psychoses or association with psychotic manifestation is unusual. We believe the diagnosis found after electroconvulsive therapy was coincidental. As there were no cerebral structural symptoms or an organic sign, MR angiography was not pursued. Takayasu arteritis should be considered in women with fever, fatigue, hypertension, and absence of pulse or low volume pulse. Early diagnosis and initiation of adequate treatment is important to prevent further progression and complications like aortic regurgitations and cerebro-vascular accidents.

Key messages

- Absent pulses in young patient should alert to possible large vessel vasculitis.
- Asymptomatic Takayasu disease is not uncommon.
- Takayasu vasculitis is entirely treatable with current newer treatment approaches.

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