

A Case of Unusual Presentation of Takayasu's Arteritis with Rheumatic Heart Disease

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Abstract: *Takayasu's arteritis is a chronic inflammatory disease of the large and medium-sized arteries. It commonly involves the aorta with its branches and the pulmonary arteries. Clinically, pulsations were absent in the involved vessels- most commonly in the subclavian artery followed by common carotid and abdominal aorta, renal artery in majority of cases. The high prevalence rates of the disease is reported from Asian countries especially in Japan, India, Korea, and Thailand. We present a 26 yrs. married female presenting with complaints of dyspnea, Paroxysmal nocturnal dyspnoea and pitting oedema over both legs since 8 months at the time of presentation. She had a past history of hemiparesis on right side about a year back which completely recovered over 4-6 hrs. On obstetrics history she had multiple spontaneous abortions (n=3) in third trimester with a preterm delivery 6 months back. On examination all pulse on right side were absent barring weaker, low volume femoral and carotid arteries. High blood pressure (200+10 and 100+10) was recorded in left brachial artery. Cardiac examination showed systolic murmur in all 4 areas. On investigation, all routine investigations were normal except a raised ESR (104). 2D echo showed severe MR/TR/PAH and moderate MS/AR (probably of a rheumatic origin). To substantiate the diagnosis of a Antiphospholipid syndrome, APLA and ANA tests were done which were negative. A CT angiography of aortic root, its branches and abdominal aorta documenting occlusions of right subclavian artery, abdominal aorta, right femoral artery and bilateral renal artery. The relative less frequently seen combination of Rheumatic heart disease in the face of a Takayasu's arteritis is presented hereby for its uncommon occurrence.*

Keywords: Takayasu's arteritis, Rheumatic heart disease, Anti-phospholipid antibody, Anti -Nuclear antibody

1. Introduction

Takayasu arteritis is a chronic non-specific inflammatory disease mainly involving the aorta and its main branches including the brachiocephalic, carotid, subclavian, vertebral, renal arteries, coronary and pulmonary arteries^{1,2}. It is defined as "granulomatous inflammation of the aorta and its major branches" by the Chapel Hill Consensus Conference on the Nomenclature of Systemic Vasculitis³. Indian origin aortoarteritis is a rare variant of this disease. There is no clear association between Takayasu's arteritis and rheumatic heart disease described in the literature. We report a case of Takayasu's arteritis in combination with rheumatic heart disease.

2. Case Report

A 26 year old female visited to Dhiraj general hospital with complain of shortness of breath and bilateral lower limb oedema since last 6 months. On history breathlessness was progressive in nature, initially it was grade MMRC grade 2 then it progressed to grade 4, associated with orthopnoea and paroxysmal nocturnal dyspnoea. She also had pitting oedema over both legs since 8 months at the time of presentation. She had a past history of hemiparesis on right side about a year back which completely recovered over 4-6 hrs. On obstetrics history, she had three multiple spontaneous abortions in third trimester and a preterm delivery 6 months back. On examination, all pulse on right side were absent barring weaker, low volume femoral and carotid arteries. High blood pressure (200+10 and 100+10) was recorded in left brachial artery.

Cardiac examination showed systolic murmur in all 4 areas. A cardiology opinion was sought and her 2D echo showed severe MR/TR/PAH and moderate MS/AR (probably of a rheumatic origin). To substantiate the diagnosis of an Antiphospholipid syndrome, APLA and ANA tests were done which were negative. Her lipid profile and coagulation profile was done, both of them were negative. On investigation, all routine investigations were normal with normal thyroid function test except a raised ESR (104).

A CT angiography of aortic root, its branches and abdominal aorta documenting occlusion of abdominal aorta, total occlusions of right subclavian artery, right superficial femoral artery, and left external iliac artery 1 cm from its origin and bilateral renal artery with multifocal ectasia of descending thoracic, infra-renal abdominal aorta and left proximal and distal subclavian artery.

3. Discussion

Over a period of 18 years (1972-1990), 83 cases of Takayasu's arteritis were reported in Indian Literature showing a female to male ratio to be 1.6:1. Sixteen per cent of the patients presented with fever and arthralgia⁴. Aortoarteritis of Indian origin is a chronic granulomatous, necrotizing vasculitis, predominantly affecting the aorta with its branches⁵. Takayasu's arteritis of Indian origin is mostly attributed to be of tuberculous, streptococcal or a collagen vascular origin. Recently more emphasis has been given on an immunopathological cause.^{6,7}

The initial vascular lesions frequently occur in the left middle or proximal subclavian artery. As the disease progresses, the left common carotid, vertebral, brachiocephalic, right middle or proximal subclavian artery, right carotid, and vertebral arteries, and aorta are also affected. The abdominal aorta and pulmonary arteries are involved in approximately 50 percent of patient.

Takayasu's arteritis has been linked to rheumatic fever and other streptococcal infections, rheumatoid arthritis, and other collagen vascular diseases⁸. In 1988, Doi et al reported a patient with Takayasu's disease and non-rheumatic mitral stenosis⁹. Nikolic et al reported a case of Takayasu's arteritis associated with aortic regurgitation and mitral stenosis¹⁰.

4. Conclusion

Takayasu's arteritis is an underreported or a rare condition reported predominantly in females. Tuberculosis and streptococcal infections are commonest causes of Takayasu's arteritis specifically in developing country like India. This patient is a classic case of Takayasu's arteritis with Rheumatic heart disease.

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