



A Case of Takayasu's Arteritis

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Case Report

A 48 year old hypertensive female presented with complaints of retrosternal chest pain with burning sensation and abdominal discomfort. On examination, patient had prominent suprasternal pulsations and absent pulses in left upper limb. Blood pressure was noted to be 150/90 mmHg in right upper limb but could not be measured on left side. Cardiovascular examination revealed a loud S₁ with no associated murmurs, a 'swishing' sound was audible over abdominal aorta suggestive of bruit. She denied any complaints of claudication pain in her limbs, joint pains or fever. Electrocardiograph done showed features of left ventricular hypertrophy and normal sinus rhythm. Chest radiograph revealed prominence of descending thoracic aorta with wall calcifications and focal dilatations [figure 1]. Laboratory investigations like hemogram, serum creatinine, ESR and CRP were normal. A CT Aortogram [figure 2] done revealed complete occlusion of left subclavian artery origin with calcific change at the origin, circumferential irregular wall thickening with circumferential calcifications [involving

inner wall] and irregular areas of stenosis seen. Focal erosions of medial wall in distal thoracic aorta were seen. Saccular aneurysm [measuring 8.0 x 7.0 x 7.7mm] arising from infra renal aorta with chronic wall changes was also noted. Severe narrowing of right renal artery with stenosis and a contracted right kidney noted [confirmed on contrast CT: figure 3]. These above mentioned findings were consistent with large vessel arteritis. A 2 dimensional echocardiography showed concentric left ventricular hypertrophy with no regional wall motion abnormality and ejection fraction of 62%.

Figure 1



Figure 2

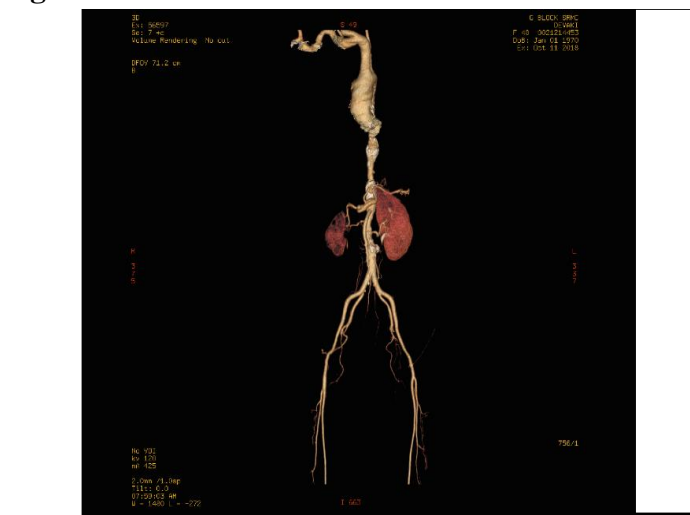


Figure 3



Takayasu arteritis (TAK) is classified as a large-vessel vasculitis because it primarily affects the aorta and its primary branches. Women are affected in 80 to 90 percent of cases, with an age

of onset that is usually between 10 and 40 years. It has a greatest prevalence in Asia. The pathogenesis is poorly understood, although cell-mediated mechanisms are thought to be of primary importance. The American College of Rheumatology (ACR) classification criteria were developed in 1990 which is used for TAK demonstrates that if 3 out of the following 6 criteria are required to establish a diagnosis. The criteria include:

Age at disease onset ≤ 40 years

Claudication of extremities

- Decreased pulsation of one or both brachial arteries
- Difference of at least 10 mmHg in systolic blood pressure between arms
- Bruit over one or both subclavian arteries or the abdominal aorta
- Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or other causes.
- Our patient fulfilled 4 of the above mentioned criteria resulting in diagnosis of TAK.

The association of renal artery stenosis resulting in secondary hypertension in patients of TAK is not uncommon. According to case report 'Takayasu arteritis in India- B.K. Sharma et al' which concluded that TAK is the commonest cause of renovascular hypertension in India.

The mainstay of therapy for Takayasu arteritis (TA) is glucocorticoids. Angioplasty, bypass grafts, or other surgery may be necessary once large aneurysms develop or irreversible arterial stenosis has occurred. Our patient was initiated on 40mg of oral prednisolone with antiplatelets. Her symptoms of retrosternal burning pain and epigastric discomfort were resolved with pantoprazole and was symptom free during her hospital stay.

Although the diagnostic criteria for TAK were long established, there are some drawbacks, so the

ACR and the European League against Rheumatism (EULAR) are in the process of making an international effort to develop revised classification criteria and diagnostic criteria.

4. Johnston SL, Lock RJ, Gompels MM. Takayasu arteritis: a review. *J Clin Pathol* 2002;55:481-6.

Learning Points

- 1) The physical examination of a patients with TAK should particularly focus on accurate measurements of blood pressure, palpation of pulses, identification of bruits, and careful cardiac auscultation.
- 2) Imaging studies are essential for establishing the diagnosis and for determining the extent of vascular involvement. Patients with suspected TAK should undergo imaging of the arterial tree by magnetic resonance angiography (MRA) or computed tomography angiography (CTA) to evaluate the arterial lumen.
- 3) Glucocorticoids are mainstay of therapy. Trial of methotrexate, cyclophosphamide, mycophenolate or other disease modifying anti rheumatic drugs can be used in steroid resistant cases. Percutaneous transluminal angioplasty or bypass grafts may be considered in late cases when irreversible arterial stenosis has occurred and when significant ischemic symptoms are present. Angioplasty is preferable

References

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