CASE REPORT

Role of 3D CT and CT Angiogram in Takayasu Aortitis (TA) of Descending Thoracic Aorta: A Case Report

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Abstract:

A 35 year old female presented with hypertension, chest and neck pain with tenderness especially towards back for several months. Pulling of face to the right, sweating of hands and lower abdomen, palpitation, joint stiffness were among other symptoms. She was suffering from occasional dyspnoea and exertional fatigue. On examination, she was found to have anaemia, hypertension, and systolic murmur. ECG was within normal limit and first echocardiography revealed an extra cardiac turbulence right to the right ventricular apex with the flow pattern suggestive of coronary artery fistula or AV malformation. She was sent for cardiac CT and CT angiogram. The result was excellent; 3D CT and CT angiogram clearly revealed a large segmental severe smooth narrowing of her descending thoracic aorta. The intra-thoracic collateral arteries were found markedly dilated including the internal mammary arteries. No VSD or coronary or AV fistula was detected. Cardiac CT also revealed moderate calcium into her heart pericardium and coronary arteries. The patient fulfilled the obligatory criteria and five minor criteria for diagnosis of Takayasu aortitis.

Introduction:

Takayasu aortitis is an inflammatory arterial disease of unknown etiology affecting aorta and its major branches with destruction of aortic and/or arterial wall and subsequent extensive replacement of the wall with fibrous tissue. Diagnosis is often difficult in early systemic (prepulseless) stage due to non-specific clinical manifestation. Disease in general takes progressive course with wax and wane of symptoms from the early inflammatory stage to the late occlusive (pulseless) stage. Early treatment may affect the prognosis of the disease.

The disease was first described on a young female with visual impairment and absent carotid pulses by Japanese ophthalmologist, Takayasu in 1908. Originally thought to occur only among Asians in young age but now known to occur in all races and all age group. Most commonly affects Asian and women in the 2nd and 3rd decades of life. About 80%-90% occur in females from age 10 to 50 years, presenting symptoms before age 30 years with mean age of 15. There is increased occurrence in monozygotic twins and in individuals with

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haplotype HLA-B5. Most often affects the elastic-type arteries with vasa vasorum such as aorta and proximal portion of its branches and also pulmonary arteries; Muscular-type arteries such as the peripheral artery are not involved.

No etiologic factor has been identified, though tuberculosis or allergic reaction to tuberculosis was once suspected. Auto-immune disease primarily affecting the aorta by both humoral and cellular immune mechanism was strongly suspected. Cellular immunity appears to play major role to induce granulomatous inflammation with T-cell help.

Case report:

The 35 year old hypertensive lady who was having Tachycardia, exertional fatigue, chest, back and neck pain and tenderness for several months but eventually not relieved by conventional symptomatic treatment. She finally came to a Cardiac surgeon with the suspected diagnosis of muscular VSD. Most of the investigations were repeated nothing new but differential diagnosis of Coronary fistula/AV malformation was added. Cardiac catheterization was advised by echo-cardiologist. Before cardiac catheterization the cardiac CT was advised. Cardiac CT was done with the following Protocol: Range-Tracheal bifurcation to heart bottom. Kv-120. mAs-700-900. Colliomation- 0.6 mm. Slice thickness- 0.75 mm. Pitch- 02 mm. Rotation time-0.33sec. Reconstruction interval- 0.5 mm. Kernel-D30f.

Procedure/Technique: High resolution ECG-Synchronized Multi-Detector Computed Tomography of the heart with particular attention to the great vessels was performed using siemens Heart View CT (64 Slice). Patient's heart rate was optimized using β-blocker and 80 ml of contrast agent (Omnipaque350) was injected in antecubital vein at 05 ml/sec followed by 50 ml saline wash.

3D CT and CT angiogram with VRT reconstruction shows severe narrowing of the descending thoracic aorta. Length of narrow segment is 33 mm and the most stenosed (>90%) luminal diameter is 2.3 mm at the centre. The wall of the involved segment is thick and irregular with signs of mild contrast enhancement. Small mural calcification was seen in the anteromedial aspect of the thickened wall. There was marked dilatation of the collaterals of the intrathoracic arteries including both internal mammary arteries. As suspected before no VSD, coronary fistula or AVM is noted. Misinterpretation of echo-cardiography as coronary artery fistula of arterio-venous malformation (AVM) may be due to prominent dilated internal mammary artery (IMA). No ventricular septal and valve defect was also noted. With one major and more then five minor criteria fulfilled the disease was finally diagnosed as a case of "Takayasu Arteritis" (Type-III) of the descending thoracic aorta with consequent dilatation of the other intra thoracic arteries with collaterals including both IMA.

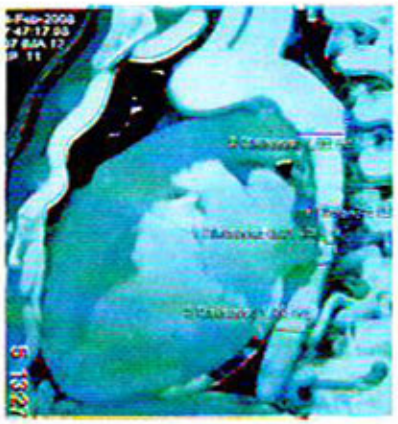

Figure-1

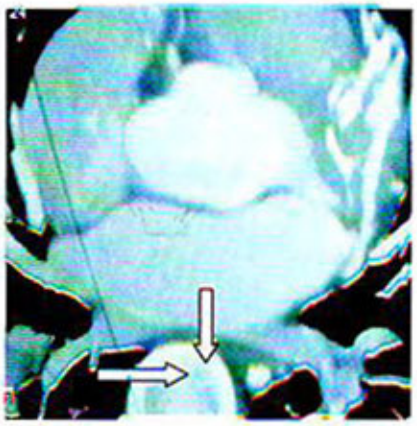

Figure-2

Figures 1 and 2: CT: Sagittal MIP images showing long segment smooth severe narrowing of descending aorta. Axial view; showing luminal narrowing (thin arrow), wall of artery and calcification (thick arrow) in the wall.

Figure-3

Figure-4

Figures 3 and 4: CT angiogram: 3D VRT images showing long segment involvement of descending aorta, heart, great vessels and other dilated intrathoracic arteries including IMA.

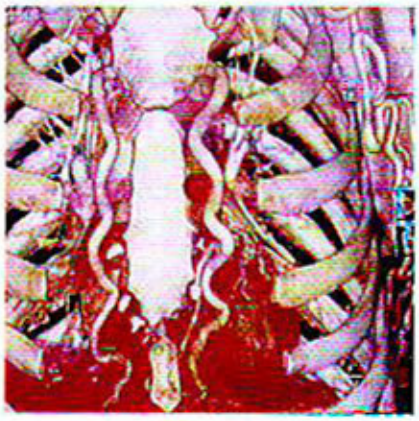

Figure-5

Figure-6

Figures 5 and 6: 3D VRT images of the grossly dilated intrathoracic arteries including both IMA.

Discussion:

Takayasu arteritis (TA) is very rare, non-specific granulomatous panarteritis of unknown aetiology most probably related to the autoimmune disease mechanism, involving entire vessel wall of elastic arteries only. TA is a world-wide disease involving all the races of all the age groups, though predominantly affecting young Asian females in the second and third decade.

The early diagnosis of TA could be made only with strong suspicion and awareness of this disease, since its symptoms as clinical manifestation are non-specific in its acute prodromal stage. The majority of TA will reach to the late (pulseless) stage with the extensive involvement of the arterial trees with wax and wane of the clinical course when the disease should progress through many years.

TA affects entire arterial wall in various forms (e.g. stenosis or dilatation) at various sites, including coronary artery and pulmonary artery besides most common involvement of ascending aorta, aortic arch and its brachiocephalic branches (e.g. carotid and subclavian arteries), descending thoracic aorta, and abdominal aorta and its branches (e.g. renal artery, mesenteric artery). Stenosis to occlusion of arterial lumen in various degrees is the most common form of the late sequellae of TA but the aneurysmal dilatation is also not uncommon in TA.

Arteriography still remains as the gold standard for the proper diagnosis and also for the surgical and endovascular management of TA as road map. MR Imaging, CT scan and Ultrasonography as non-invasive diagnostic tests, are essential for the proper early diagnosis of TA by the selective assessment of acute and chronic changes of the intra-mural components of the vessel wall before the arteriographic study is able to confirm the intraluminal changes of TA in its later stage. Non-invasive study is also essential for the assessment of the response to the treatment of TA besides the follow-up assessment of the progress of the disease for the long-term care.

Systemic medical treatment with steroid and/or immunosuppressive therapy in early acute stage of TA is critical with good response in general. Surgical (operative) treatment based on bypass operation and/or endovascular (interventional) treatment based on percutaneous transluminar angioplasty (PTA) with or without stent insertion can improve overall results of the therapy in the late occlusive, non-inflammatory, quiescent stage.

Fully integrated medical, operative and interventional therapies through multidisciplinary approach can improve the overall success rate of the treatment of TA either in acute or chronic, or active or inactive stage of the disease with reduced morbidity in long term care of TA.

Advanced non-invasive diagnostic modalities like 3D CT with CT angiogram and advanced MR Imaging can provide more precise information of the intramural status of the vessel wall during the early acute and chronic active phase of the disease for the proper timely diagnosis and management to improve the treatment result with reduced over-all morbidity. Multidisciplinary approach to integrate the various medical, operative and endovascular treatment modalities can improve overall results of long term care through its acute and chronic stages altogether. Thus CT including CT angiogram and 3D CT with VRT reconstruction alone can play a great role in early diagnosis of the disease with stages. Post-operative evaluation and follow up can also be done with modern CT.

References:

1. Johnson SL, Lock RJ, Gompels MM. Takayasu's arteritis: A review. J Clin Path 2002; 55: 481-486
2. Mehra NK, Jaini R, Balamurugan A, et al. Immunogenetic analysis of Takayasu arteritis in Indian patients. Int J Cardiol 1998; 66 (Suppl 1): S127-32
3. Lupi-Herrera E, Sanchez-Torres G, Marcushamer J, Mispireta J, Horwitz S, Vela JE. Takayasu's arteritis. Clinical study of 107 cases. Am Heart J 1977; 93: 94-103
4. Subramanyan JH, Joy J, Balakrishnan KG. Natural history of aortoarteritis (takayasu's disease). Circulation 1980; 10: 429-37
5. Amano J, Suzuki A. Coronary artery involvement in Takayasu's arteritis; collective review and guidelines for surgical treatment. J Thorac Cardiovasc Surg 2003; 125: 570-7
6. Jain S, Kumari S, Ganguly NK, Sharma BK. Current status of Takayasu arteritis in India. Int J Cardiol 1996; 54 Suppl: S111-6
7. Sharma S, Kamalakar T, Rajani m, Talwar KK, Shrivastava S. The incidence and pattern of pulmonary artery involvement in Takayasu's arteritis. Clinic Radiol 1990; 42: 177-82
8. Ishikawa K. Natural history and classification of occlusive thromboaortopathy (Takayasu's disease). Circulation 1978; 57: 27-35.