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Spectrum of CT Angiographic Findings in a Series of 114 Patients of Clinically Suspected Cases of Takayasu Arteritis – A Prospective Study at a Teritiary Care Hospital in India

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Abstract

Aims & Objectives: *1) To detect various angiographic patterns by CT angiography in clinically suspected 114 cases of Takayasu's arteritis.2)To characterize & classify the lesions into various subgroups 3) To evaluate diagnostic & clinical usefulness of CT angiographic cross sectional imaging for establishing the diagnosis of Takayasu's Arteritis 4) To discuss the differential diagnosis.*

Summary: *In our study group, the age of clinical onset of disease varied from 7 yrs to 50 yrs with female preponderance. The commonest site of involvement in our study is Abdominal aorta & the second common site was Renal artery. The younger age group of patients showed more extensive involvement & in multiple vessels*

Conclusion: *CT angiography is one the most reliable ,relatively commonly available method of investigating Takayasu arteritis as it can provide reliable information regarding the mural & luminal abnormalities of aorta & its branches & pulmonary artery*

Keywords: *Takayasu's arteritis, CT angiography.*

Introduction

The condition is named after Dr Takayasu (Japanese ophthalmologist) who initially described similar vascular features on the retina in 1908. Takayasu arteritis is a chronic idiopathic inflammatory disease that primarily affects large vessels such as aorta & its branches & pulmonary artery & coronary artery [(1)]. The disease shows female preponderance with majority of patients in 20 -30 yrs age groups. Though the etiology is unknown, many theories have been postulated on etiological factors like auto immune mediated, genetic predisposition & heriditory causes. Histopathologically, Early Takayasu Arteritis changes consist of an adventitial mononuclear infiltrate with perivascular cuffing of the vasa vasorum followed by medullary mononuclear inflammation occasionally accompanied by granulomatous changes [(7)] Two forms have been described Japanese & Indian forms. In Japanese

population, association with BW 52, DR 12 antigens is found in 50 % population but not seen in other countries. In Japan ,ascending aorta, arch & its branches are more commonly affected therefore aortic regurgitation is more common Whereas in Korea ,India &western countries ,abdominal aorta and its branches & renal arteries are more affected therefore hypertension & cerebrovascular accidents are more common. Halotype of HLA -A-24-B -52 - DR -2 is frequently found in cases of aortoarteritis with inflammatory abdominal aortic aneurysms which are more prone to accelerated progression of inflammation which is refractory to antiinflammatory therapy. For example, in Japan and Korea there is a clear association with the extended haplotype: HLA B*52, DRB1*1502, DRB5*0102, DQA1*0103, DQB1*0601, DPA1 *02-DPB1*0901.[11–14] Takayasu arteritis has been associated with different human leucocyte antigen (HLA) alleles in different populations [11,12] .

In the present day, Common modalities of investi-gations are Colour Doppler, CT angiography, MR angiography, Nuclear medicine imaging, PET -CT & Digital substraction angiography. The advantages of CT Angiography are that it is non invasive, accurate assessment of size & extent of lesion, relationship to other anatomical structures, perianeurysmal hematoma due to leaking aneurysms can be better evaluated, visceral vessels & peripheral vessel evaluation is more accurate than MRA ,can be done even if breath holding capacity is limited. The disadvantages being that higher radiation dosage is required for the procedure & use of nephrotoxic contrast agents.

Materials & Methods

Study Period: March 2010 to May 2016 (6 years)

Source of the Data: The patients admitted in Gandhi hospital with clinical suspicion of Takayasu's arteritis referred to the department of Radiodiagnosis for CT angiography evaluation.

Study Equipment: CT was performed on SOMATOM EMOTION 16 SLICE (SIEMENS Ltd)

Study Sample: 114 cases of clinically suspected aorto arteritis

Exclusion criteria: 30 cases were showing normal findings on angiography but showing positive findings for other causes of hypertension like cardiac, suprarenal pathologies etc & 14 cases were showing diffuse atherom-atous changes were all discarded from the study. 70 cases showed diffuse or focal areas of vascular pathologies

Clinical Presentation: [7] Takayasu Arteritis has traditionally been divided into an early, "prepulseless" systemic phase, and a late, occlusive phase. In the early systemic phase, diagnosis is difficult and symptoms are usually nonspecific and constitutional, including fever, myalgias, weight loss, and arthralgias. In the occlusive phase, ischemic symptoms dominate, including angina, claudication, syncope, and visual impairment. Late-phase TA may be further subclassified as classic pulseless disease (type 1), a mixed type (type 2), an atypical coarctation type (type 3), and a dilated type (type 4). Most patients present with a form of late-phase disease.

CLASSIFICATION

There are many methods of classifications in Takayasu Arteritis

ACR CRITERIA FOR CLASSIFICATION OF TAKAYASU'S ARTERITIS: A diagnosis of Takayasu's arteritis requires that at least 3 of the 6 criterias are met.

Sr. No.	Criteria
1	Age at disease onset ≤ 40 years.
2	Claudication of extremities.
3	Decreased brachial artery pulse.
4	Blood pressure difference > 10 mm Hg.
5	Bruit over subclavian arteries or aorta.
6	Arteriogram abnormality.

1. According to the type of lesion morph-ology: it is classified into stenosing ,deforming & aneurysmatic types

2. ANGIOGRAPHIC CLASSIFICATION OF TAKAYASU'S ARTERITIS (TAKAYASU CONFERENCE, 1994) [2]

Type	Vessel involvement
Type I	Branches from the aortic arch
Type II a	Ascending aorta, aortic arch, and its branches
Type II b	Ascending aorta, aortic arch and its branches, thoracic descending aorta.
Type III	Thoracic descending aorta, abdominal aorta,and/or renal arteries.
Type IV	Abdominal aorta and/or renal arteries.
Type V	Combined features of Type II b and IV.

3.ISHIKAWA CLASSIFICATION: Based on presence & severity of complications,

Table 2: Ishikawa clinical classification of Takayasu arteritis [10,11]

Group	**Clinical features**
Group I	Uncomplicated disease, with or without pulmonary artery involvement
Group IIA	Mild/moderate single complication together with uncomplicated disease
Group IIB	Severe single complication together with uncomplicated disease
Group III	Two or more complications together with uncomplicated disease

Diagnostic Criteria

CLINICAL PRESENTATION: [7] Takayasu Arteritis has traditionally been divided into an early, "prepulseless" systemic phase, and a late, occlusive phase. In the early systemic phase, diagnosis is difficult and symptoms are usually nonspecific and constitutional, including fever, myalgias, weight loss, and arthralgias. In the occlusive phase, ischemic symptoms dominate, including angina, claudication, syncope, and visual impairment. Late-phase TA may be further subclassified as classic pulseless disease (type 1), a mixed type (type 2), an atypical coarctation type (type 3), and a dilated type (type 4). Most patients present with a form of late-phase disease.

CT Angiographic features of Takayasu Arteritis

CT Angiography remains the gold standard for diagnosis of Takayasu Arteritis. Assessment of pulmonary vasculature by angiography is not universally recommended, being reserved for patients with symptoms of pulmonary hypertension[9,11] Curved planar reformation (CPR) allows tortuous vessels to be displayed along its long axis; multiplanar reconstruction (MPR) gives the anatomical information of arteries in the optimal planes; volume-rendered (VR) images can illustrate the extension of the luminal lesions and map the collaterals following artery occlusion. A combination of CPR, MPR, VR, and axial images permits optimal evaluation of luminal changes [1].

The most common vascular changes described are a) Those occurring in the vessel wall: wall thickening, wall edema, wall enhancement b) In the lumen: stenosis, occlusion, aneurysm formation 3) Presence of collaterals

Mural Changes: The typical changes that occur in the wall of vessels are concentric mural thickening which can be of varying degrees focal to several millimeters in length. On pre-contrast CT scanning, the mural thickening is of high attenuation compared with the lumen, while on the post-enhanced CTA images, it exhibits a double ring enhancement pattern, which is typically shown in venous phase [3,6,1] It has been proposed that the inside ring represents the swollen intima, while the outside ring indicates the active inflammation in the medial and adventitial layers [6,1].

Luminal changes: Luminal Stenosis is frequently associated with mural thickening seen both in major vessels & in branch vessels. Commonly affected vessels are abdominal aorta, descending aorta, subclavian artery, common carotid artery & renal arteries. Aneurysms are more common in abdominal aorta, descending aorta which may lead to aortic rupture. Perianeurysmal hematoma due to leaking aneurysms can be better evaluated by CT Angiography than any other investigative modality & its early & accurate diagnosis prevents fatal outcome

Presence of Collaterals: Evaluating for the presence of collaterals helps in planning treatment protocols along with assessment of extent of luminal narrowing in the parent vessel.

Maximum intensity projection and VR are useful in demonstrating small vessel changes

Discussion

This is a prospective study of 114 patients of clinically suspected cases of Takayasu Arteritis referred to Dept of Radiodiagnosis, Gandhi Hospital for CT angiography. CT angiography was performed on SOMATOM EMOTION 16 SLICE (SIEMENS Ltd)with 3D reconstruction . The Study period was from March 2010 to May 2016 (6 years). The Diagnosis & Sub group classification were done based on CT angiograp-hic findings in correlation with clinical findings.

In the present study group of 114 patients in whom CT angiography procedures performed, 30 cases were showing normal findings for angiography but showing positive findings for other causes of hypertension like cardiac, suprarenal pathologies etc & 14 cases were showing diffuse Atheromatous changes were all discarded from the study.70 cases showed diffuse or focal areas of vascular pathologies. 70 cases of clinically suspected aorto arteritis with hypertension were screened by CT angiography with 3D reconstuction. Most the patients presented with variable pulselessness, variable blood pressure recordings in different limbs. Diagnosis is always delayed because of nonspecific symptoms in acute phase but clues for diagnosis are hypertension, vascular bruits, assymetrical arm blood pressure. Despite the disease being recognized for more than 100 years, the outlook for patients with takayasu arteritis remains poor and the treatment suboptimal [8]

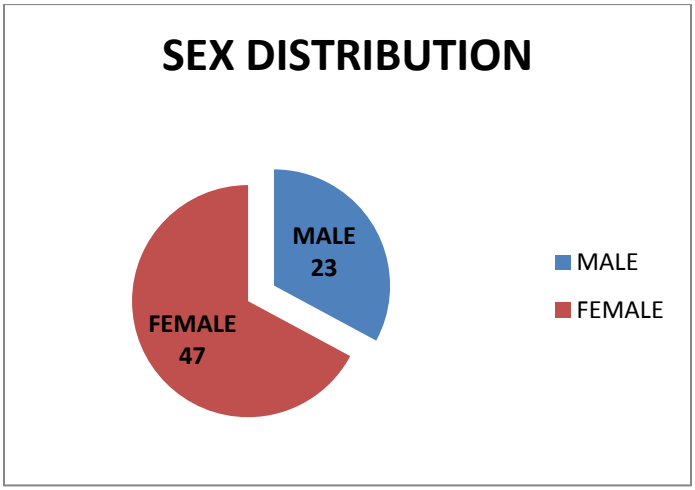
In the present study group of 70 patients, Females were predominantly affected (67.14 %) than male patients (32.85 %) with peak age group in 21 -30 yrs. The youngest patient in the age group was 7 yrs old & oldest was 47 yrs of age . Most of the patients with clinical onset of disease were between 7 to 50 years with female prepnderance and female to male ratio being 2: 1 approximately In a study done by Linu Cherian Kuruvilla [8], The peak age group with female preponderance coincided with that of our study however the Ratio of female patients were much higher in their study probably due to smaller sample group.

Clinical symptoms

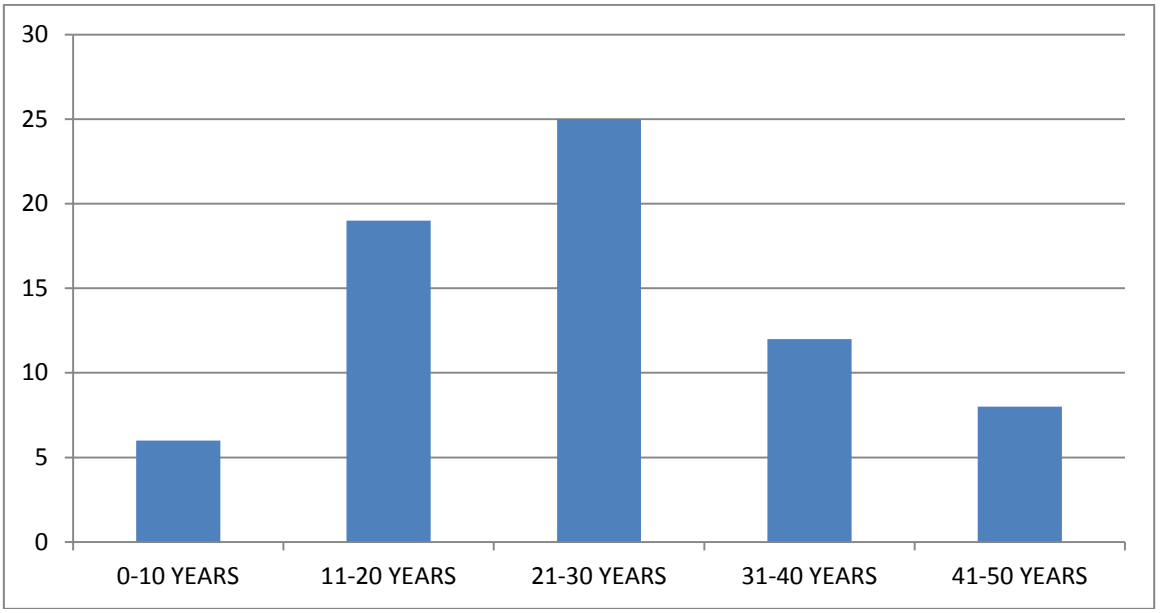
Table 1

Symtoms	No:of Patients	Percentage (%)
Hypertension	24	34.28 %
Stroke in young	15	21.42 %
Tingling & pain in extremities	3	4.2%
Persistant headache not responding to medications	8	11.42%
Headache with giddiness & vomiting	12	17.14%
Pulselessness	15	21.42%
Fever with body pains	8	11.42 %
Multiple vascular bruits	15	21.42 %

In the present study group, hypertension was the commonest symptom (34.28%) followed by pulselessness, multiple vascular bruits & stroke in young (21.42 %)



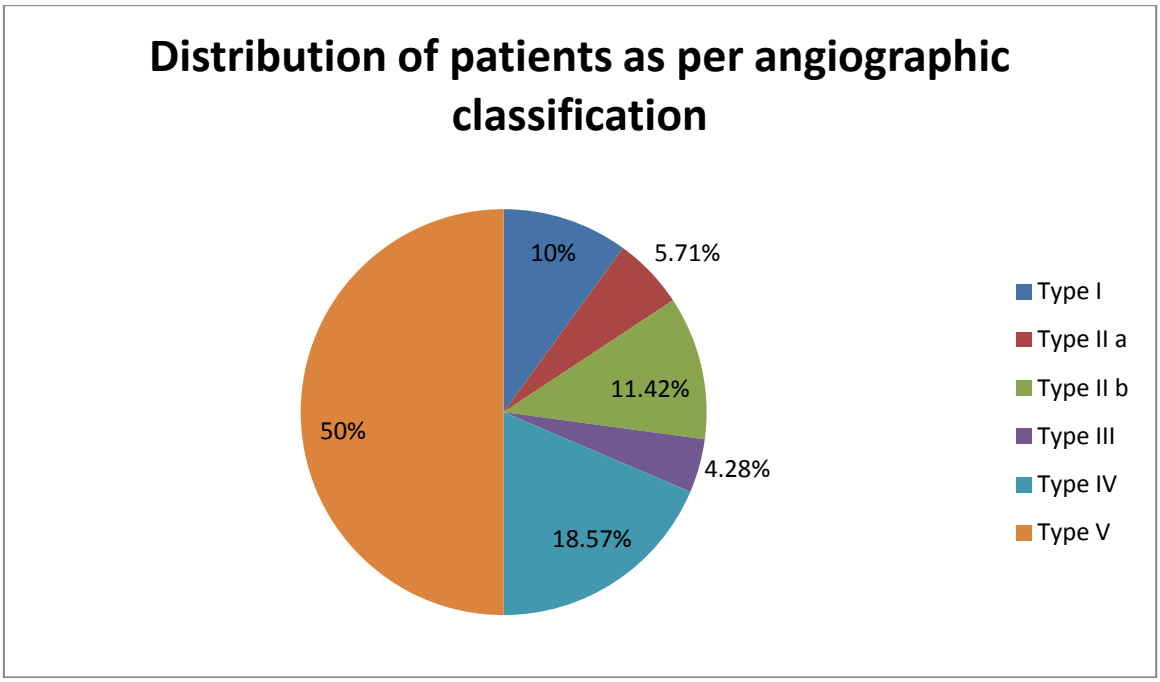
PIC 1: represents the sex distribution in our study. Out of 70 cases, 47 (67.14%) were female patients & 23 (32.85 %) were male patients



Pic 2: represents the age (of onset of clinical symptoms) distribution of patients in our study. The number of patients were 6 (8.57 %) in 0-10yrs, 19 (27.14 %)in 11- 20 yrs age group, 25 (35.71 %) in 21 – 30 yrs age group , 12 (17 . 14 %) in 31 – 40 yrs & 8 (11. 42%) in 41-50 yrs.

Table 2 : Represents the distribution of patients as per angiographic classification

TYPE	No:of patients	Percentage
Type I	7	10 %
Type IIa	4	5.71%
Type IIb	8	11.42%
Type III	3	4.28%
Type IV	13	18.57%
Type V	35	50%



Pic 3: represents the distribution of patients in our study according to angiographic classification in this study as per angiographic classification most of the patients were in Type V followed by Type IV both accounting for 68. 57 %.

Differential Diagnosis: a) Atherosclerosis: calcified plaques are more common. Atherosclerotic plaques are more common in patients aged 45 yrs or above, not usually associated with long segment stenosis [3]

b) Gaint cell arteritis: both Gaint cell arteritis & Takayasu arteritis can cause destruction of vessel wall, commonly affects large vessels & characteristically show the presence of gaint cells but Takayasu arteritis shows characteristic skip lesions affecting both Intima & Adventia of the vessel. In Gaint cell arteritis branches of external & internal carotids are more commonly affected

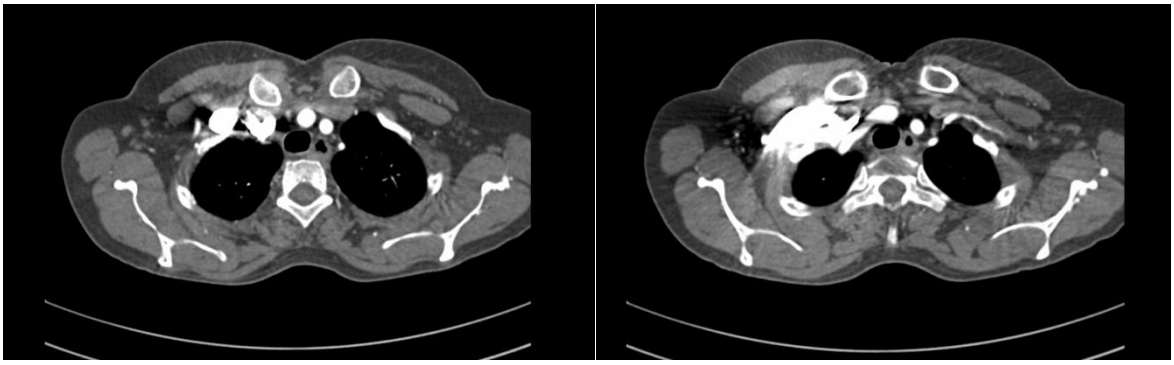
c) Polyarteritis Nodosa : frequently occurs in adults who are 30–50 years old, affecting males more than females, and it also more commonly affects patients with hepatitis B. Gastrointestinal and renal arteries are the primary sites diseased. Multiple small aneurysm formation in the involved artery is the characteristic manifestation on CTA images [1,5]

d) **Tuberculous Aortitis:** tends to cause erosion of the vessel wall with the formation of true or false aneurysms, particularly affecting the descending thoracic and abdominal aorta. Dissection and rupture are important complications rather than the stenoses typical of Takayasu arteritis [11]

COMPLICATIONS

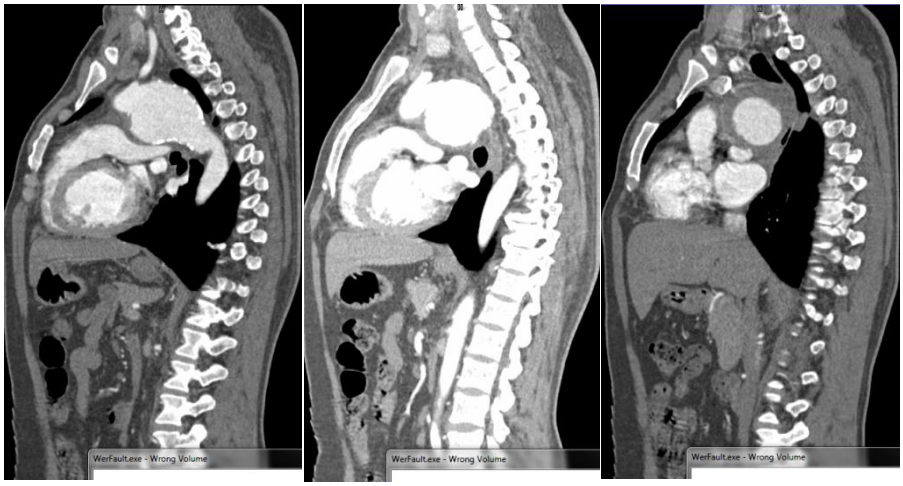
The four most important complications were defined as Takayasu retinopathy, secondary hypertension, aortic regurgitation, and aneurysm formation, each being graded as mild / moderate or severe at the time of diagnosis [10,11]

CASE 1



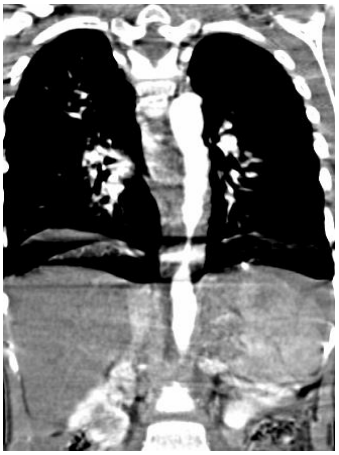
TYPE I : Reduced calibre of left sub clavian artery, partial stenosis of right subclavian at the ostium.

CASE 2



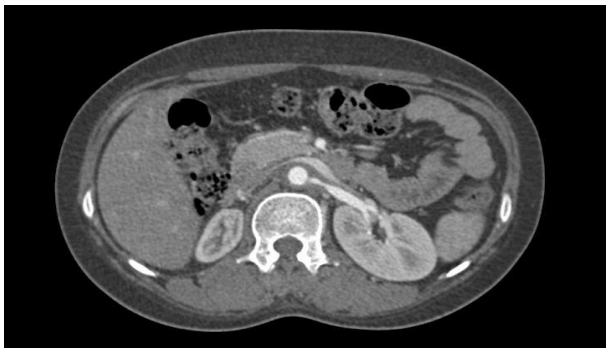
TYPE II b: Dilatation of arch of aorta upto proximal descending aorta. Mural thrombus in the aneurysm. Calcified aneurysmal wall.

CASE 3



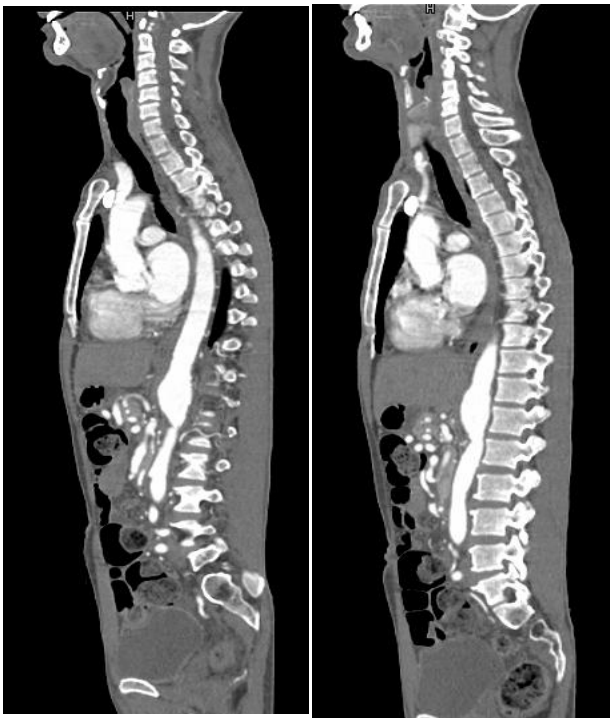
TYPE III: Narrowing of distal thoracic aorta.

CASE 4



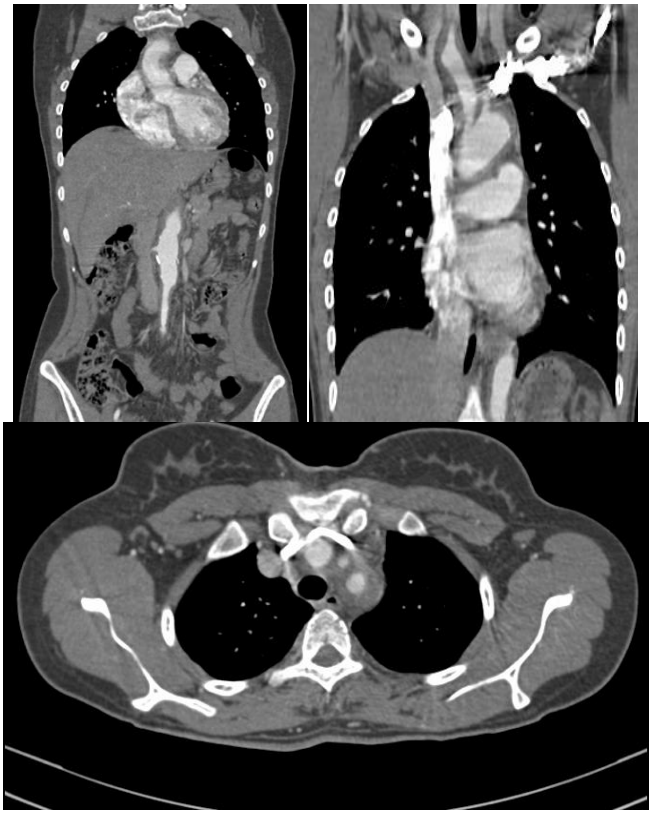
TYPE IV: Narrowing of the left renal artery from the origin and reduced calibre of abdominal aorta.

CASE: 5



TYPE IV: Aneurysmal dilatation of proximal abdominal aorta.

CASE 6



TYPE V: Partial luminal narrowing with wall thickening of distal abdominal aorta, left common carotid artery at the origin.

Conflict of Interest: Nil
Source of Funding: Nil

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