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The Many Faces of Takayasu Arteritis with Digital Subtraction Angiography- A case series.

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Abstract:
Background: Takayasu arteritis is a rare, systemic chronic inflammatory large vessel vasculitis that causes obliterative stenosis and occlusion and post-inflammatory dilatation in arteries. It comprises of varied clinical features and imaging spectrum due to number of vessel involvement. Symptoms are usually non-specific. Digital Subtraction Angiography is considered the gold standard to evaluate location and type of arterial involvement.
Objectives: The aim of this study was to determine the clinical and imaging spectrum of Takayasu's arteritis and to classify the disease based on angiographic findings.
Material and methods: The study was conducted at the Department of Radio-diagnosis, Medical College Kolkata. A total of thirty patients with suspected Takayasu arteritis were evaluated. The study was conducted between December 2020 to December 2021.They were further evaluated with colour doppler sonography and digital subtraction angiography. The data was analysed and compared in terms of age of onset, gender predisposition, clinical features, type and location of arterial involvement and classified accordingly.
Results: In our study we found that the age group 30 to 45yrs were more commonly involved. Females are more commonly affected (90%) than males. Maximum patients complained of pain in upper limb (33%). Most commonly involved artery was subclavian artery (90%). Stenotic lesions were more common than occlusive or dilatation. Majority of the patients were of type I and I and IV combined.
Conclusion: Digital subtraction angiography plays an important role in defining the type and location of arterial involvement in the form of stenosis, occlusion, dilatation as well as guide appropriate therapeutic options and follow-up.
Keywords: *Takayasu arteritis, Digital Subtraction Angiography, Stenosis, Occlusion, Artery.*

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I. Introduction

Takayasu arteritis (TA) is a rare, systemic, idiopathic granulomatous large-to-medium vessel arteritis that predominantly involves the aorta, its major branch arteries, and (less frequently) the pulmonary arteries **[1].** The disease has been referred to with a number of different names in the past, such as aortic arch syndrome, pulseless disease, idiopathic aortitis, stenosing aortitis, aorto-arteritis, and occlusive thrombo-arteriopathy. The worldwide incidence is estimated to be 2.6 cases per million per year **[2]**. There is strong female predominance in the ratio of 9:1, affecting mainly the Asian populations **[3]**. The typical age of presentation is around third decade. Takayasu arteritis is associated with substantial morbidity and mortality. In only 20% cases TA is monophasic and self-limited. But in majority of cases, it is progressive and requires immunosuppressive treatment. So early diagnosis and treatment is necessary to alter the disease outcome and prevent future complications. Patho-physiology reveals infiltration of mononuclear macrophages and lymphocytes in tunica media. It causes stenotic, occlusive or aneurysmal change in the large to medium sized arteries. Destruction of tunica media leads to aneurysmal changes. Renal arteries may also be involved leading to hypertension.

II. Materials And Methods

Thirty patients with clinically suspected Takayasu arteritis were referred to our department of Radiodiagnosis in our tertiary care hospital Medical College, Kolkata and Hospital. Proper history taking, clinical examinations, laboratory investigations and imaging studies including colour Doppler sonography (with GE Logitech P9, Philips iu22 ultrasound machine) & digital subtraction angiography (with Allura xper fd20

machine) were conducted during the study period of one year from August 2020 to August 2021.The patients who had a history of contrast sensitivity were excluded from the study. The patients were studied and analysed descriptively in terms of age group, gender predisposition, clinical features, percentage of artery involvement, type of artery involvement and classified accordingly.

III. Results

Case 1- A 38-year-old female was referred due to tingling sensation and pain in right upper limb. Doppler ultrasound demonstrates reversed flow in left vertebral artery as compared to common carotid artery (left subclavian steal phenomenon) and circumferential symmetric thickening of lumen of common carotid artery **(Figure 1).** Digital Subtraction Angiography (DSA) was done. It showed-

1.Left subclavian artery occlusion from its origin with reformation by Left vertebral artery exhibiting "Vertebral Steal Phenomenon" **(Figure 2,4).**
2.Right subclavian artery occlusion with reformation of Right axillary artery through collaterals **(Figure 3).**
3. An intra-vascular stent measuring 2.5 cm is noted at the origin of left subclavian artery without any flow. S/O-restenosis or non-functioning stent **(Figure 4).**
4.Bilateral common carotid artery stenosis at the level of bifurcation.
5.Right external carotid artery shows gross stenosis (70%) at its origin from the CCA.
6.Left external carotid artery shows moderate stenosis (50%) at its origin from the CCA.

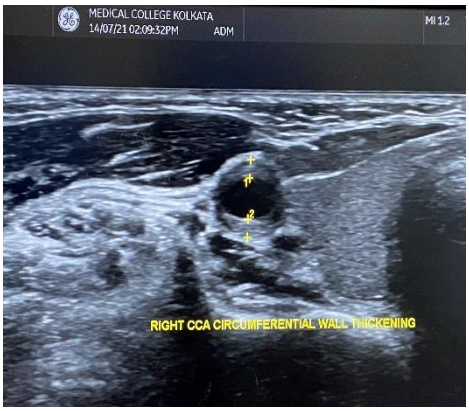


Figure 1:Symmetric circumferential wall thickening of right common carotid artery.

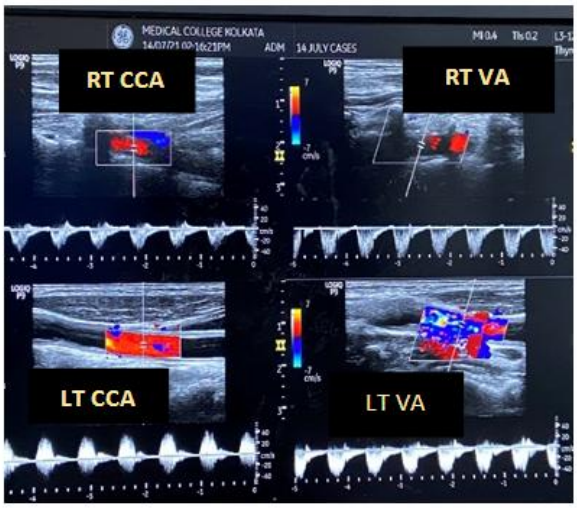


Figure 2:Reversal of waveform of left vertebral artery compared to left common carotid artery (Vertebral Steal Phenomenon). Normal waveforms in right side.

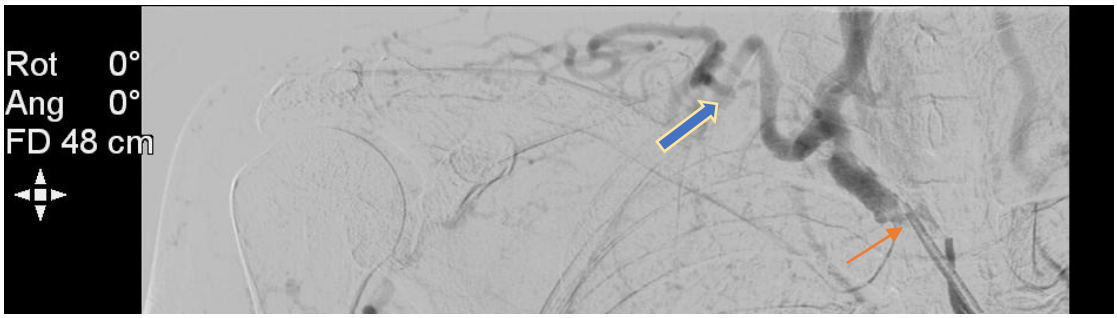


Figure 3:Right Subclavian artery occlusion (orange arrow) and reformation of right axillary artery by collaterals (blue arrow).

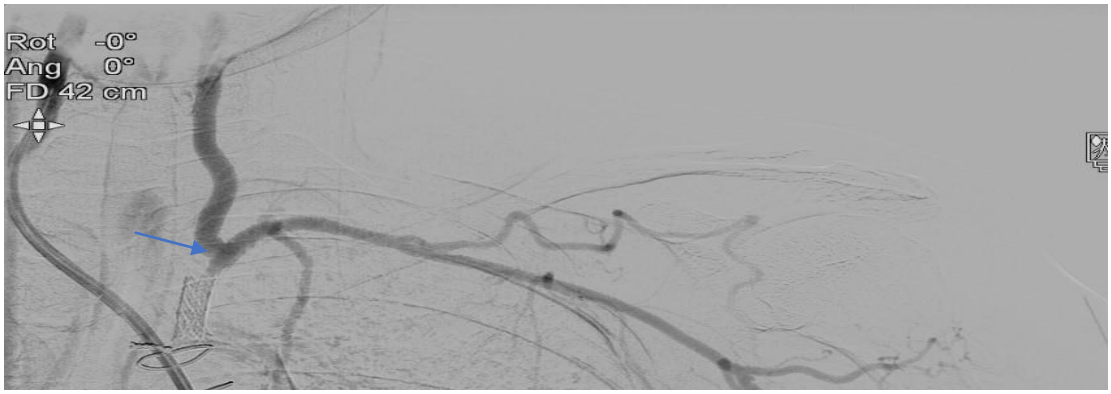


Figure 4: Left subclavian artery occlusion at its origin and reformation by left vertebral artery (blue arrow). Stent noted in situ.

Case 2-

A 32-year-old female was referred due to lower limb claudication. Doppler ultrasound reveals stenosis of the lumen of the lower abdominal aorta. DSA demonstrates-

1. Occlusion of abdominal aorta at the level of L3 **(Figure 5).**
2. Occlusion of left axillary artery.
3. Anterior tibial, posterior tibial, peroneal artery of both lower limbs are narrowed in calibre **(Figure 6).**
4. Bilateral dorsal pedis artery, medial and lateral calcaneal artery, arcuate artery and plantar artery could not be demonstrated.

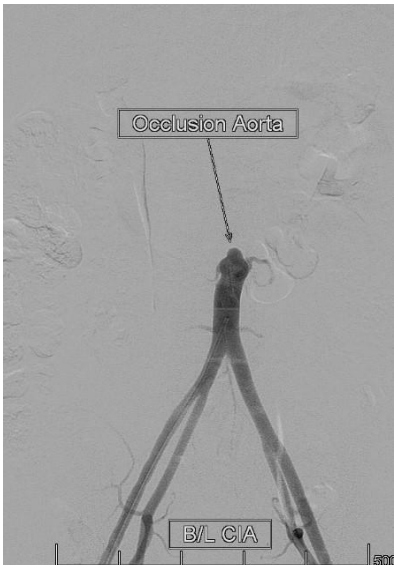


Figure 5:Occlusion of abdominal aorta at the level of L3.

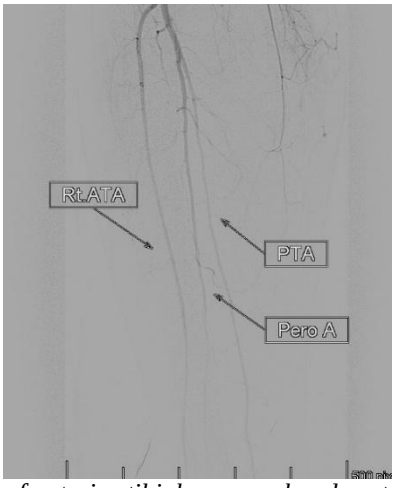


Figure 6:Stenosis of anterior tibial, peroneal and posterior tibial artery.

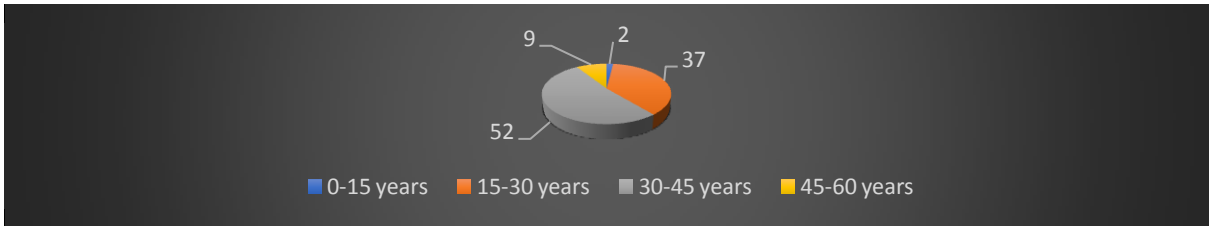


Table 1:To illustrate the age distribution of patients.

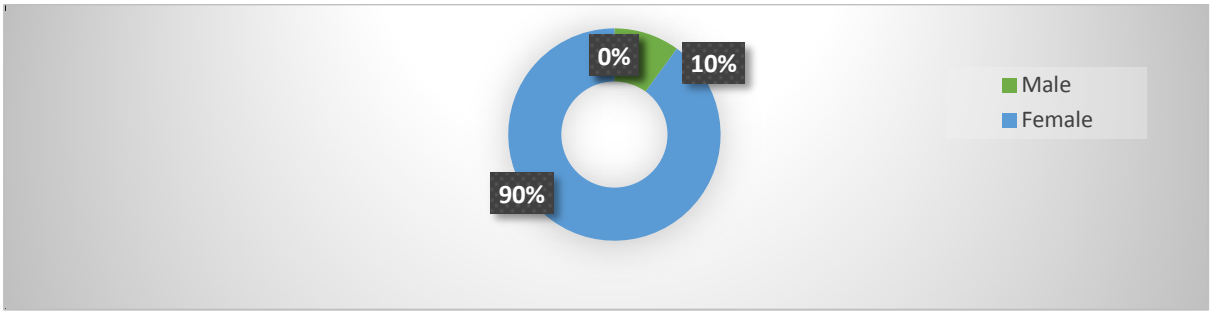


Table 2:To illustrate the gender predisposition of the subjects.

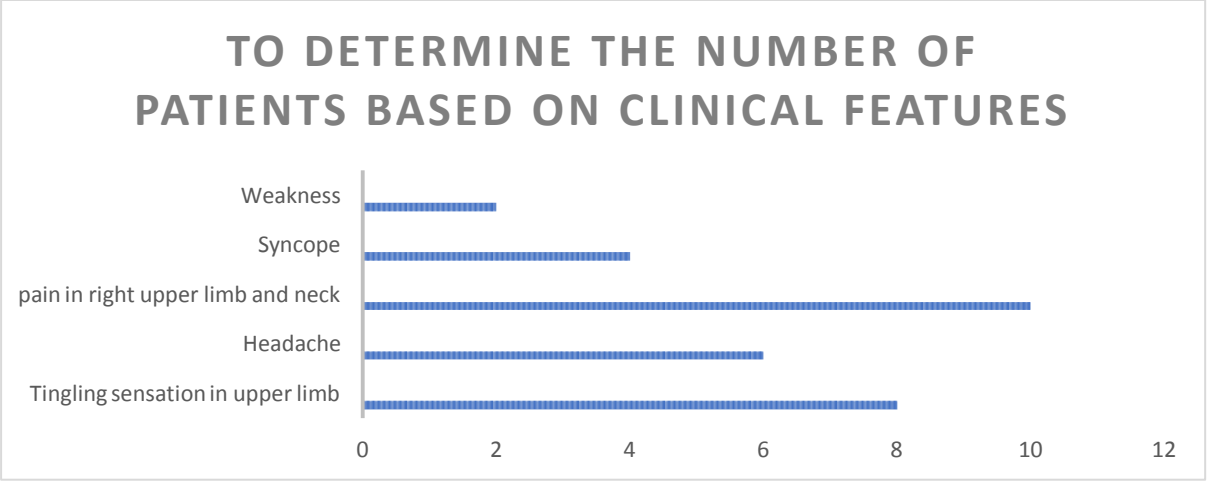


Table 3:To determine the number of patients based on the clinical features.

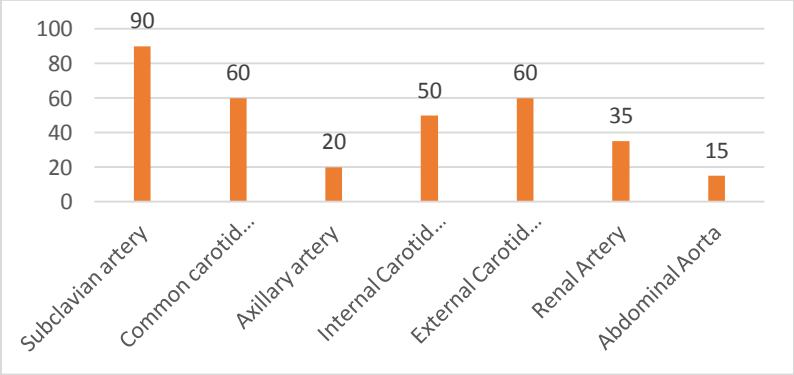
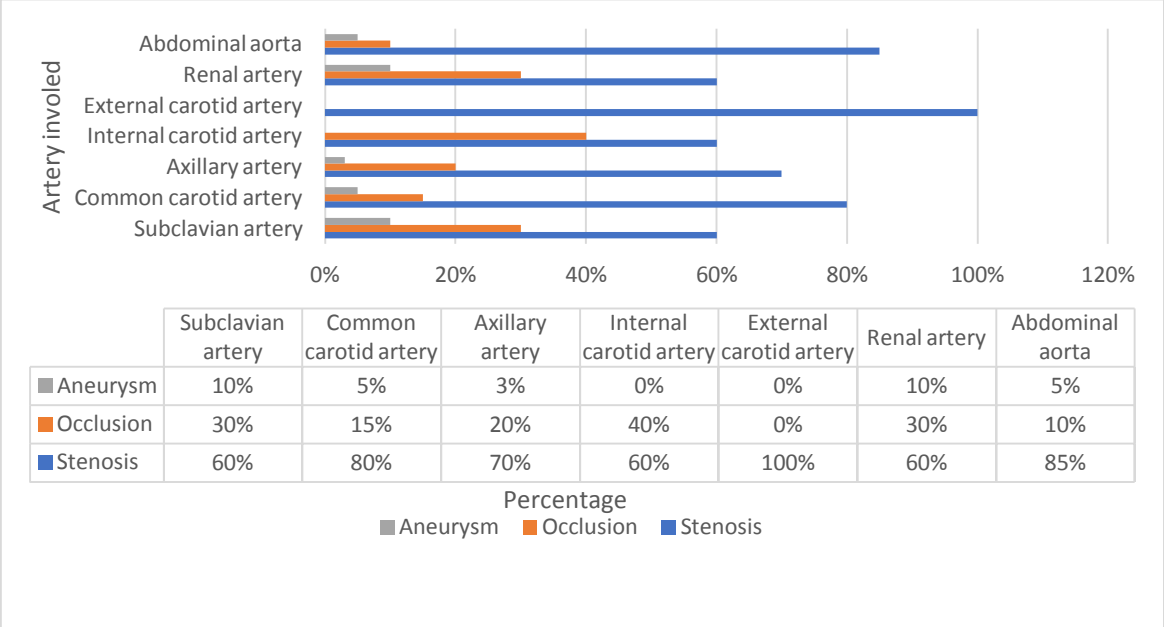


Table 4:To determine the percentage artery involvement in the study.



	Subclavian artery	Common carotid artery	Axillary artery	Internal carotid artery	External carotid artery	Renal artery	Abdominal aorta
Aneurysm	10%	5%	3%	0%	0%	10%	5%
Occlusion	30%	15%	20%	40%	0%	30%	10%
Stenosis	60%	80%	70%	60%	100%	60%	85%

Table 5: Type of arterial involvement in the study.

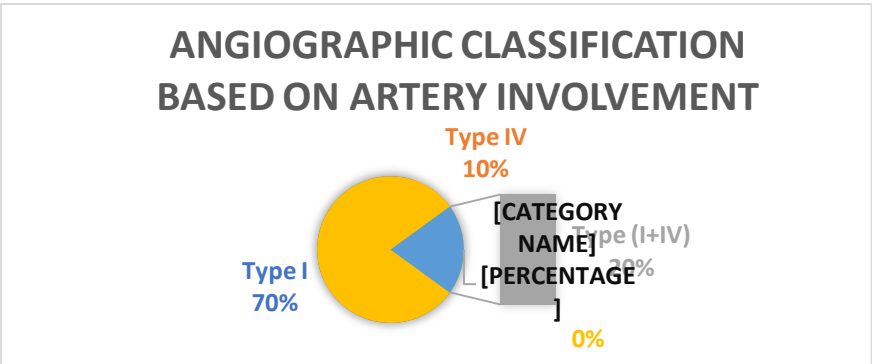


Table 6: Angiographic classification based on the artery involvement

IV. Discussion

In our study of 30 patients during the period from December 2020 to December 2021 we found that the age group 30 to 45yrs were more commonly involved **(Table 1).** Females are more commonly affected than males in the ratio 9:1 **(Table 2).** Maximum patients complained of pain in upper limb (33%) **(Table 3).** The other clinical features were episodes of syncope, weakness in limbs, headache and tingling sensation in upper limb. These features were related to the location of artery involved. Hypertension was associated with renal artery involvement, syncope with carotid artery involvement and upper limb symptoms were related to the subclavian artery involvement. The following arteries were involved in descending order; subclavian arteries (90% of total cases), common carotids (60% of total cases), renal arteries (35% of total cases) and abdominal

aorta (15% of total cases) **(Table 4).** Stenosis was more common than complete occlusion in all arteries **(Table 5).** Majority of the patients were of type I (64%), type IV (9%) whereas 27 % belonged to type I+IV combined **(Table 6).**

The first case of Takayasu arteritis was described in 1908 by a Japanese ophthalmologist Mikito Takayasu at the annual meeting of Japan Ophthalmology Society, where Takayasu described a peculiar "wreathlike" appearance of the blood vessels in the back of the retina **[5].** The disease progression is said to occur in a biphasic pattern. Initially, in the pre-pulseless systemic phase, the diagnosis is difficult as it generally causes non-specific constitutional symptoms like malaise, fever, fatigue, weight loss and myalgia. Pulseless phase is the late occlusive phase where there is obliterative granulomatous changes in the arteries causing stenosis, occlusion and aneurysm creating a myriad of presentations like reduced or absent peripheral pulses, renovascular hypertension, limb claudication and other features of limb ischaemia, angina, syncope & visual impairment **[3]**. The etiology of Takayasu arteritis is unknown. The underlying pathologic process is inflammatory with several etiologic factors having been proposed including infection with Spirochetes, M. Tuberculosis and circulating auto antibodies due to autoimmune process **[1].**

There is segmental granulomatous inflammation of arterial wall with marked intimal proliferation and fibrosis of media and adventitia leading to stenosis & occlusion. Occasionally post stenotic dilatation and aneurysm formation may result when the vascular media is damaged by inflammatory process. These changes lead to many complications e.g., hypertension due to renal artery stenosis, aortic insufficiency due to aortic valve involvement and pulmonary arterial hypertension leading to interstitial pulmonary fibrosis, congestive cardiac failure. The other complications include ischemic retinopathy, vertebrobasilar ischemic microaneurysm, carotid stenosis, hypertensive encephalopathy and inflammatory bowel disease. Rarely, Takayasu arteritis has been associated with glomerulonephritis and ankylosing spondylosis **[4,6].**

Diagnosis of Takayasu arteritis comprises of clinical suspicion, laboratory investigations (raised ESR and CRP) and imaging modalities. No serological marker available so clinician has to rely on imaging for diagnosis and accurate classification. The imaging modalities are-

1.Ultrasonography- Long segment symmetrical homogeneous thickening of arterial wall also known as "Macaroni Sign" is highly specific for TA **[8].** In contrast, atherosclerotic plaques are calcified, involves short segment and appear with irregularly thickened walls.

2. Computed Tomography- demonstrates mural thickening and enhancement with intramural calcification **[9].**

3. Magnetic Resonance Imaging- ideal for follow-up and staging of patients. Contrast enhanced MRI can demonstrate concentric thickening and hyperenhancement of inflamed arterial wall and mural thrombosis.

4.Digital Subtraction Angiography (DSA)- Although considered the gold standard, other non- invasive imaging methods like colour doppler sonography, CT angiography, MR angiography are becoming more popular nowadays **[10].** However multiple long segment stenosis ranging from mild to severe to complete occlusions with collateral formation or subclavian steal phenomenon is best delineated on DSA. A characteristic finding is presence of skip lesions. Besides, therapeutic angioplasty and stent placement can be done. It is however, an invasive procedure which may result in several complications associated with large dose of iodinated contrast media radiation dose. It is difficult to evaluate the walls of the artery involved. It only gives information about the lumen of vessels.

Classification is based on location of arterial involvement **[4].**

- Type I- classic type involving the aortic arch branches namely the subclavian arteries, carotid and the brachiocephalic trunk.
- Type II a) involvement of the ascending aorta and aortic arch branches.
- Type II b) involvement of the ascending aorta, aortic arch branches and thoracic descending aorta.
- Type III - involvement of both the thoracic and abdominal aorta with or without involving the renal arteries.
- Type IV - involvement of the abdominal aorta with or without involving the renal arteries.
- Type V - Generalised involvement of all the aortic segments.

Treatment consists of systemic corticosteroids with judicious use of angioplasty and bypass procedures.

V. Conclusion

TA is a type of chronic form of large vessel segmental vasculitis resulting in stenotic and occlusive lesions involving arterial walls leading to a highly variable spectrum of presentation, varying stages of disease activity & response to treatment. Digital subtraction angiography plays an important role in defining the type and location of arterial involvement in the form of stenosis, occlusion, dilatation as well as guide appropriate therapeutic options like angiography with stenting in the inactive phase. It is also used for to monitor disease progression, remission and to check efficacy of interventional therapeutic modalities.

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