IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 17, Issue 4 Ver. 10 (April. 2018), PP 52-61
www.iosrjournals.org

An Original Article : Spectrum of Imaging Findings of Takayasu Arteritis With Digital Subtraction Angiography : A Prospective Study In A Tertiary Care Hospital

Dr. Debashis Dakshit, Dr. Aditi De, Dr. Amrit Chattopadhyay
Associate Professor, Department Of Radio-Diagnosis, Medical College, Kolkata
Junior Resident, MD Radio-Diagnosis, Medical College, Kolkata
, Junior Resident, MD Pediatrics, Calcutta National Medical College, Kolkata
**Corresponding author: Dr. Debashis Dakshit,*

Abstract
BACKGROUND- *Angiographic demonstration of the extent of involvement of large vessels in suspected cases of Takayasu arteritis.*
OBJECTIVES- *Our objective is to identify the spectrum of clinical manifestations, extent of arterial involvement and to classify the type of Takayasu arteritis based on angiographic findings.*
METHODS- *We investigated 10 patients having symptoms related to suspected Takayasu arteritis with laboratory investigations & imaging modalities like Color Doppler Sonography & Digital Subtraction Angiography in our institution, Medical College, Kolkata, (a tertiary care hospital) for a period of 1 year from January, 2017 to January, 2018. We compared the clinical characteristics of patients with angiographic findings of arterial involvement and classified the types of Takayasu arteritis according to the pattern of vascular involvement, the incidence of disease in males & females, age of onset etc.*
RESULTS- *We found that Takayasu arteritis is commoner in females, in the age group 31 to 45yrs. Approximately 60% of cases have symptoms related to upper limb claudication, 30% cases present with hypertension, headache and syncope. Among arterial involvement the subclavian arteries are involved (70%) in most cases with stenosis more common than complete occlusion & Types I & IV are more prevalent than other types.*
CONCLUSIONS- *Takayasu arteritis is a chronic form of systemic large vessel vasculitis resulting in stenotic, occlusive lesions involving arterial walls leading to a highly variable spectrum of presentation, stage, disease activity, prognosis and response to treatment.*
Keywords: *Takayasu arteritis (TA) / aorto-arteritis/ pulseless disease/ granulomatous arteritis/ classification/ interventional radiology/ digital subtraction angiography/ macaroni sign.*

Date of Submission: 02-04-2018 Date of acceptance: 17-04-2018

I. Introduction

Takayasu arteritis is a rare disorder of unknown etiology. The worldwide incidence is estimated to be 2.6 cases per million per year[^(1)] and approximately 80% are females[^(2)] of child bearing age group. The typical age of presentation is around third decade. Takayasu arteritis is associated with substantial morbidity and mortality. In 20% cases TA is monophasic and self limited. In majority of cases it is progressive or relapsing / remitting and requires immunosuppressive treatment. So early diagnosis and treatment is necessary to alter the disease outcome and prevent future complications. Takayasu arteritis has a predilection for the aortic arch and its branches. It is also known by many names such as "Aortic arch syndrome", "Non specific aorto-arteritis", "Idiopathic medial aortopathy", "Pulseless disease". However, renal arteries may also be involved leading to hypertension. It can cause stenotic, occlusive or aneurysmal change in the large to medium sized arteries. The chronic course of the disease results in irregular massive intimal fibrosis leading to narrowing of vascular lumen. Destruction of tunica media leads to aneurysmal changes. Pathophysiology shows infiltration of mononuclear macrophages and lymphocytes in tunica media.

II. Materials & Methods

Ten patients with myriads of non specific symptoms were investigated in Medical College, Kolkata with history taking, clinical examinations, laboratory investigations, imaging studies including color Doppler sonography (Clear vue 550, HD-7, iu-22) & digital subtraction angiography (Allura xper fd20), during the period from January,2017 to January,2018. Out of the ten patients investigated, eight of them were females and

two males. Amogst the various spectrum of manifestations, the most common presenting symptom was tingling sensation in the upper limbs associated with diminished radial pulse with laboratory work up revealing raised ESR and CRP. Other clinical presentations were nonspecific like, inability to carry out normal daily activities, neck pain, syncope, headache, shortness of breath.

The angiographic findings of arterial involvement were corroborated with clinical history and Takayasu arteritis was classified according to the angiographic characteristics of vascular lesions.

III. Results

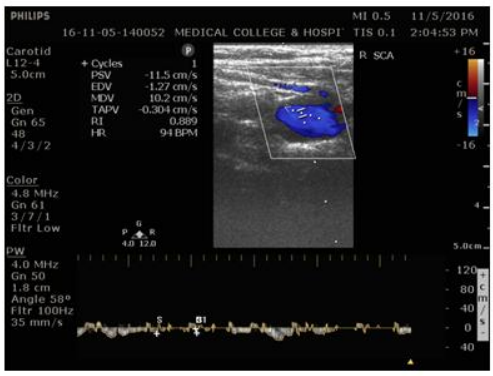


Fig.1a Right subclavian artery – reduced flow

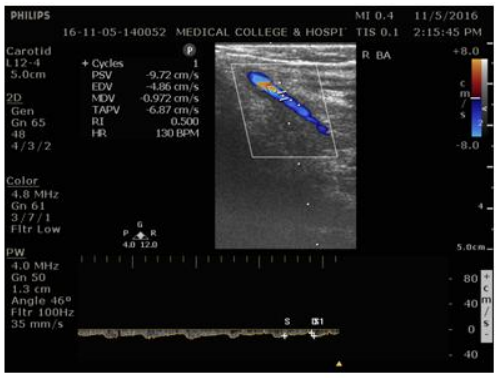


Fig.1b Right brachial artery - reduced flow

Doppler ultrasound of upper limb arteries, in a 19 yrs/F showing low flow in right subclavian and brachial arteries.

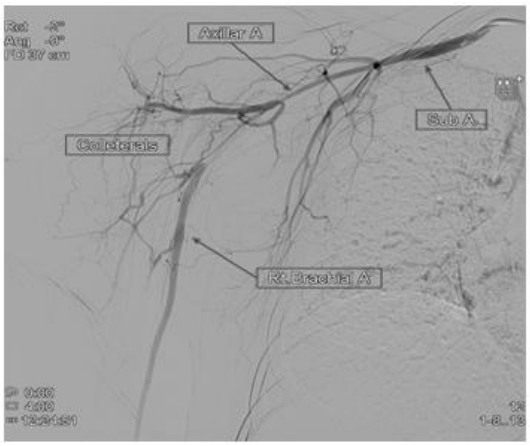
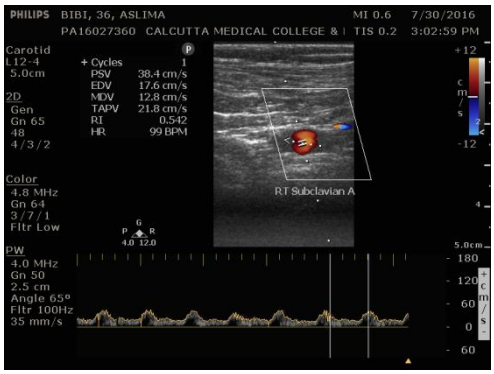


Fig.1c Right upper limb DSA

DSA in the same patient reveals narrowed distal subclavian, axillary and brachial arteries which are seen reformed distally by collaterals.The color Doppler ultrasound in a 36 yrs female patient, shows complete flow void signal in the right common carotid artery. The right subclavian artery shows reduced PSV with a biphasic spectral pattern & the right axillary and brachial arteries show gradual reduction in flow with monophasic spectral pattern.



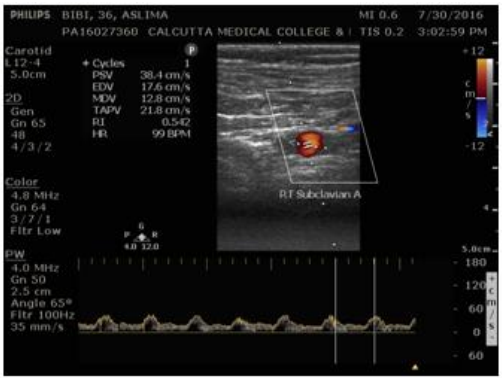
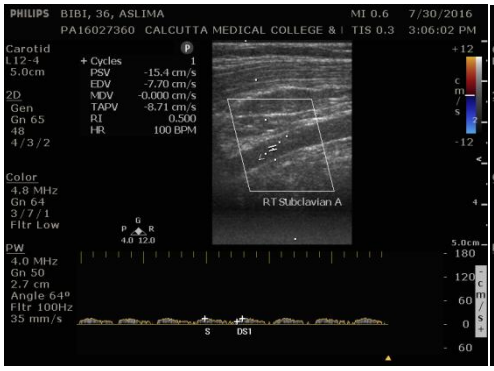


Fig: 2a Right common carotid artery-no color flow **Fig: 2b** Right subclavian artery(proximally)



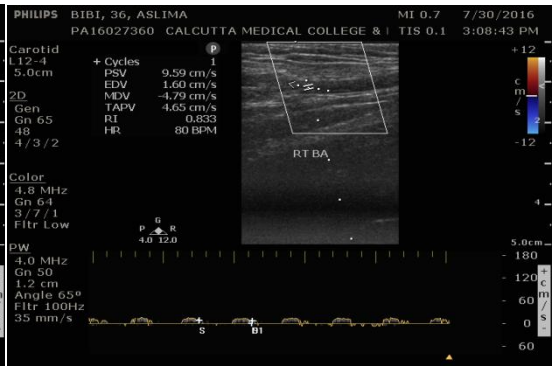


Fig: 2c Right subclavian artery (distally) **Fig:2d** Right brachial artery

DSA in the same patient, reveals proximal narrowing of right CCA and right SCA. The right axillary artery was reformed by collaterals.

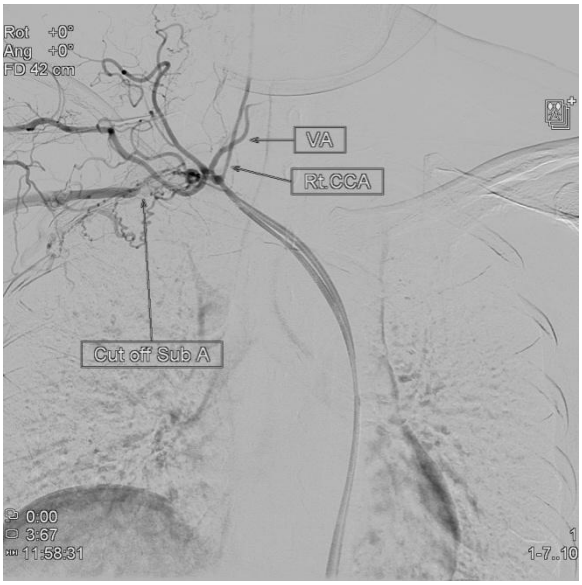


Fig.2e Right upper limb DSA

In a 40yr old male patient, with sudden onset loss of consciousness and severe headache, GCS – 8/15, BP – 184/72 mm Hg in right upper limb and 84/56 mm Hg in left upper limb. NCCT brain revealed intraventricular haemorrhage involving bilateral lateral ventricles.

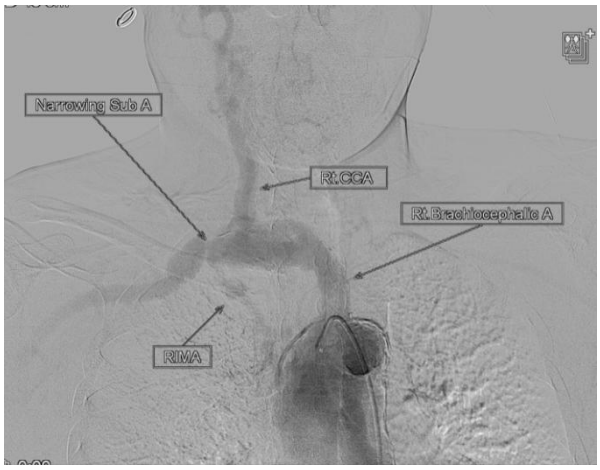


Fig.3a Aorta and it's brabches-angiography

Digital subtraction angiography reveals non-visualization of left CCA, ICA, ECA due to occlusion at the origin.

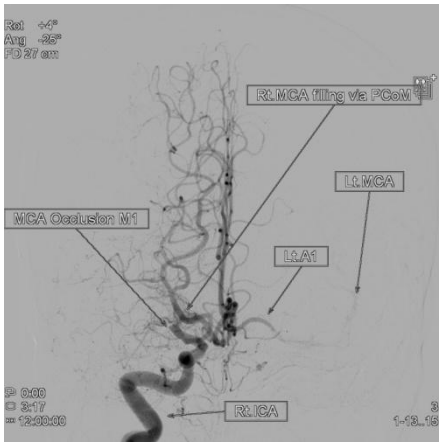


Fig.3b Cerebral angiography

Complete occlusion of right MCA at its M1 segment and filling via posterior intercommunicating artery from right PCA is seen.

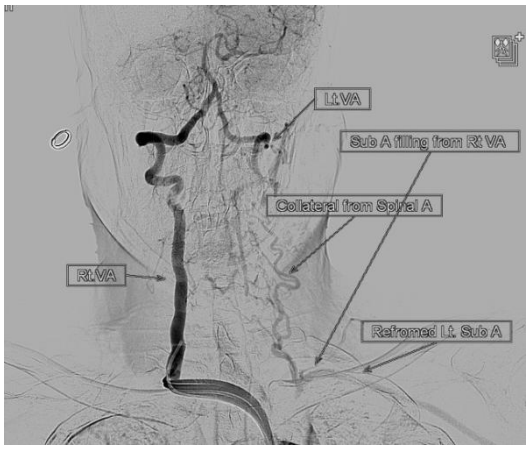


Fig.3c Cerebral angiography

Retrograde filling of left vertebral artery by flow from the right vertebral artery. Left subclavian artery is occluded from it's aortic origin at the aorta, upto the origin of left vertebral artery, distally reformed by collaterals from branches of left vertebral artery and continued as the axillary artery.

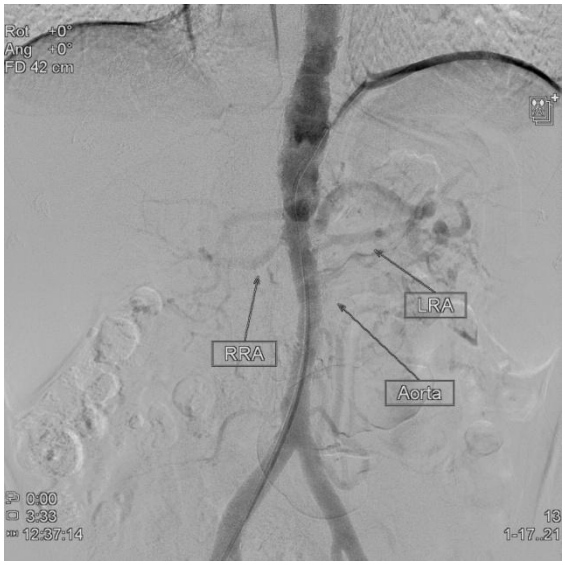


Fig.3d Abdominal aorta angiography

Abdominal aorta shows dilated, tortuous course, the infrarenal part gradually narrowed till bifurcation. Right and left renal arteries are stenosed at their origin with calibre reduction of 75% and 90% respectively.

Table - 1

	Case1	Case2	Case3	Case4	Case5	Case6	Case7	Case8	Case9	Case10
Age	19yrs	36yrs	42yrs	36yrs	38yrs	40yrs	19yrs	20yrs	38yrs	33yrs
Sex	F	F	F	F	F	M	F	F	M	F
Presenting complaints	Tingling sensation RUL, neck pain.	Headache, neck pain, low grade fever.	Tingling sensation & pain RUL, cold extrimities.	Claudication of all 4 limbs and intermittent low grade fever.	Syncopal attacks and neck pain.	Sudden onset LOC and severe headache.	Tingling sensation LUL	Headache	Weakness LUL	Syncopal attacks
BP(RUL) mm Hg	88/64	92/66	92/68	100/76	100/72	176/94	120/80	172/90	156/84	96/68
BP(LUL) mm Hg	108/74	120/82	130/88	118/78	114/80	144/90	102/74	136/88	102/70	118/72
Vessels involved	Bilaterally narrowed SCA and BA	narrowing of Rt. CCA &SCA	complete cut off of Rt. SCA at origin.	occlusion of Rt. SCA at origin, with distal collaterals. Stenosis of Lt. SCA at origin, reformed	Bilateral SCA narrowed at origin; Left distal ATA narrowed.	Complete occlusion of left CCA, ICA, ECA, right MCA at its M1 segment. Narrowing of	narrowing of left SCA at origin, with distal collaterals.	narrowing of left SCA at origin with distal collaterals. Occlusion of left axillar	Narrowing of left CCA at commencement, occlusion of left SCA	Narrowing of b/l CCA at origin and occlusion of right SCA just after origin.

				distally.		bilateral SCA.		y artery.		
Renal artery Involvement	Nil	Nil	Nil	Nil	Nil	Bilateral renal artery stenosis. (Rt. 75% Lt. 90%)	nil	Bilateral renal artery stenosis	Right renal artery stenosis	nil
Abdominal aorta involvement	Nil	Nil	Nil	Nil	Nil	Narrowing of infrarenal abdominal aorta	Nil	Nil	Nil	Nil
Angiographic classification	Type I	Type I	Type I	Type I	Type I	Type I + IV	Type I	Type I+ IV	Type I + IV	Type I

Table 2: Angiographic findings of arterial lesions in aorta & it's branches – number of arteries involved with stenosis/occlusion:

Artery	Stenosis	Occlusion
Right Common carotid	2	0
Left common carotid	1	2
Right subclavian	5	2
Left subclavian	5	2
Abdominal aorta (infrarenal)	1	0
Right renal artery	3	0
Left renal artery	2	0

Table 3 : Stenotic & occlusive lesions :

Pattern of arterial involvement	**Number of lesions**	**Percentage**
Stenosis	**19**	**70.4%**
Occlusion	**6**	**29.6%**
Total	**25**	**100%**

Chart - 1 Percentage of stenotic and occlusive lesions

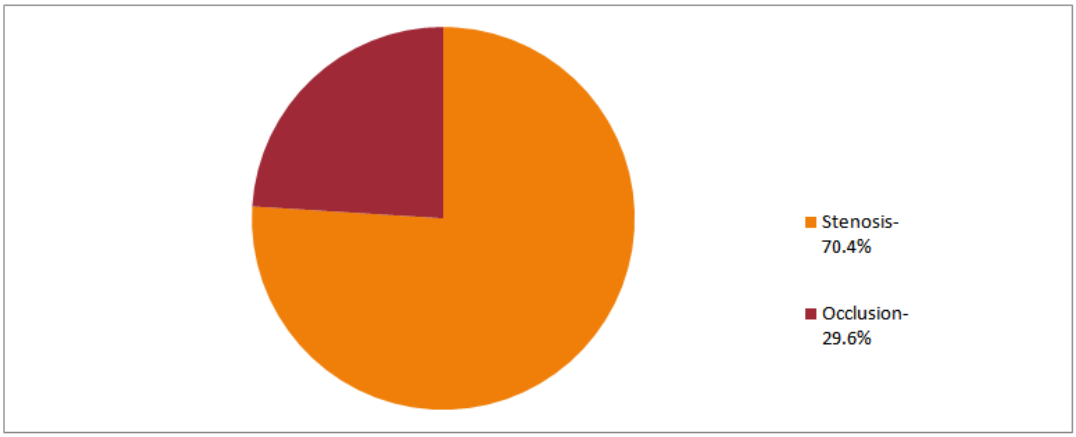


Table 4: Different arterial involvement in respect to number of cases:

Arterial involvement (stenosis, occlusion)	**Percentage in respect to total number of cases**
Right subclavian	**70%**
Left subclavian	**70%**
Bilateral subclavian	40%
Right common carotid	20%
Left common carotid	**30%**

Bilateral common carotid	10%
Right renal artery	**30%**
Left renal artery	20%
Bilateral renal artery	20%
Abdominal aorta	10%

Chart - 2

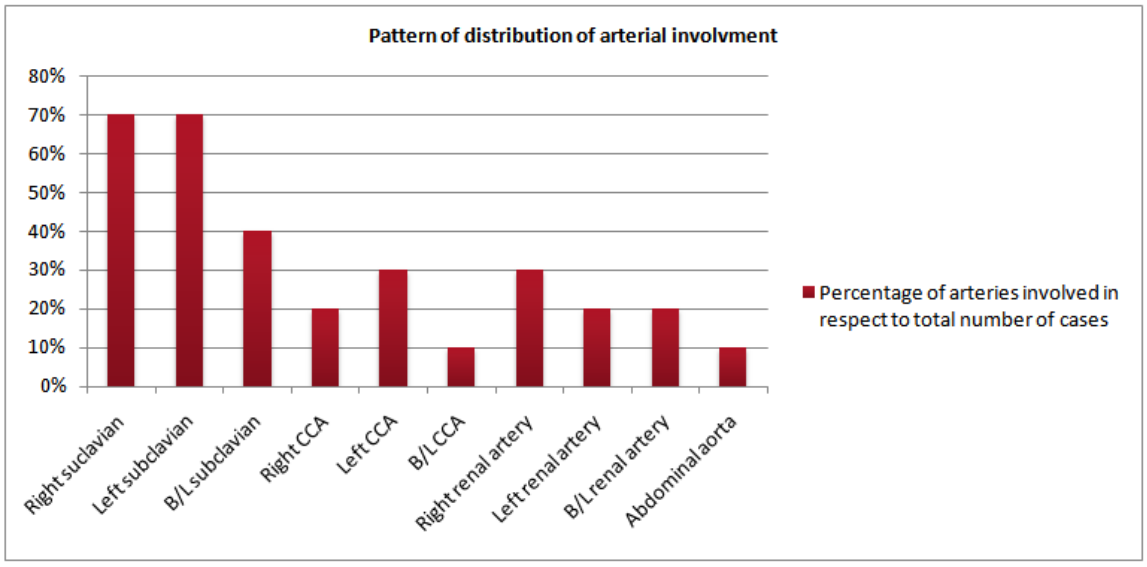


Chart - 3

Sex distribution

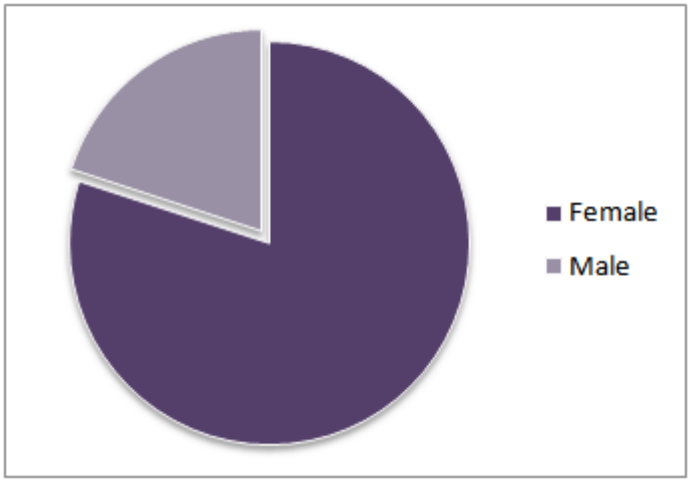


Table – 5: Sex distribution

Sex	Male	Female
Number	2	8
Percentage	20%	80%

Chart - 4
Age distribution

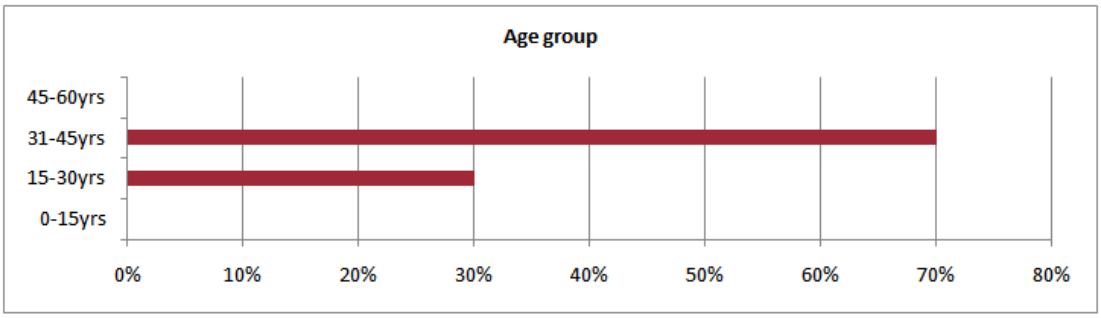


Table - 5

Age group	Number of pateints	Percentage
0-15yrs	**0**	**0%**
15-30yrs	**3**	**30%**
31-45yrs	**7**	**70%**
45-60yrs	**0**	**0%**

Chart – 5 Classification-Takayasu arteritis

Classification-Takayasu arteritis

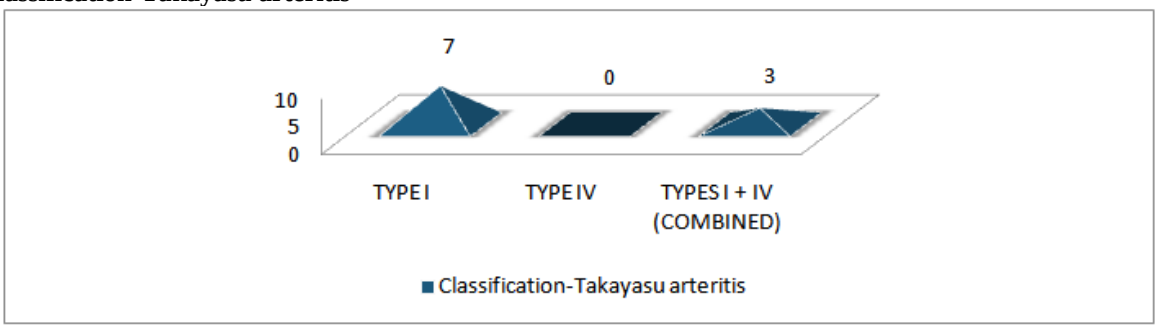


Table - 6

Types	Number of cases
I	**7**
IV	**0**
I+IV	**3**

In our study of 10 patients, during the period from January,2017 to January,2018. we found that females in the age group 31 to 45yrs were more commonly involved. The following arteries were involved in descending order; subclavian arteries (70%), common carotids (30%), renal arteries (30%) and abdominal aorta (10%). Stenosis (70.4%) was more common than complete occlusion (29.6%). Type I & combined types I & IV were more prevalent. Approximately 60% of cases had symptoms related to upper limb claudication, while 30% cases presented with hypertension, headache and syncope. The clinical spectrum of upper limb symptoms were corroborative with involvement of the subclavian arteries, syncope and headaches were common with carotid arterial disease, whereas renal artery involvement was associated with hypertension. Significant difference of blood pressure was noted between both the upper limbs.

IV. Discussion

The first case of Takayasu arteritis was described in 1908 by a Japanesse ophthalmologist Mikito Takayasu at the annual meeting of Japan Ophthalmology Soceity, where Takayasu described a peculiar 'wreathlike' appearance of the blood vessels in the back of the retina.[3]

Takayasu arteritis also known as "Pulseless disease" is a granulomatous large vessel vasculitis which predominantly affects the Aorta and its major branches.[4]

The disease progression is said to occur in a triphasic pattern.[5] Initially, in the pulseless systemic phase, the diagnosis is difficult as it generally causes non specific constitutional symptoms like malaise, fever, fatigue, weight loss, myalgia.

Phase II is the vasculitic stage where constitutional symptoms are associated with features of vascular involvement like tenderness or pain over vessels (angiodynia).

In the late occlusive phase, the presentation is usually angina, claudication, syncope & visual impairment.

The histopathological findings include Granulomatous inflammation of arterial wall with marked intimal proliferation and fibrosis of media and adventitia leading to stenosis & occlusion. Occaisionally post stenotic dilatation and aneurysm formation may result when the vascular media is damaged by inflammatory process.

Vascular changes lead to many complications e.g. hypertension due to renal artery stenosis, aortic insufficiency due to aortic valve involvement; pulmonary arterial hypertension leading to interstitial pulmonary fibrosis, congestive cardiac failure. The other complications include ischemic retinopathy, vertebrobasilar ischemic microaneurysm, carotid stenosis, hypertencive encephalopathy and inflammatory bowel disease.

Rarely, Takayasu arteritis has been associated with glomerulonephritis, ankylosing spondylosis.

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 like glomerulonephritis, ankylosing spondylosis. Genetic factors, HLA B*52 association may play a role in pathogenesis.[^(1)]

Diagnosis of TA is based on clinical information, laboratory evaluation (raised ESR & CRP) and diagnostic imaging. No definite serologic marker is available for diagnosis. Therefore, radiological features are essential for accurate diagnosis and disease classification. The imaging modalities for diagnosing TA includes –

ULTRASONOGRAPHY- Long segment smooth homogenous, echogenic circumferential thickening of arterial wall. On transverse section this is known as the 'macaroni sign' and is highly specific for TA.[^(6)] In contrast, atherosclerotic plaques appear inhomogenous, often calcified with irregular arterial walls and generally affects a short segment. Vascular occlusion due to intimal thickening and/or secondary thrombus formation, aneurysm can be seen. Contrast enhanced ultrasound identifies neovascularization on the adventitial side of arterial wall in the early stage. It is also used in treatment response evaluation.

CT – Demonstrates mural thickening with intramural calcification.[^(7)]

MRI – Ideal for follow up of patients under treatment. Contrast enhanced MRI may demonstrate concentric or cresentic arterial wall thickening, delayed hyper enhancement of the inflamed arterial walls & mural thrombosis can be easily seen. New MR contrast agent Gadofoveset trisodium enhances the arterial wall significantly in active phase of the disease. Staging of disease is important for the timing of vascular intervention as re-stenosis is a frequent complication if intervention is done in active phase.

PETCT – Involved segments show linear uptake of FDG characteristic of active vasculitis. It is an excellent technique with a high sensitivity & specificity.

DSA – Although DSA is considered the gold standard for diagnosis of TA, non-invasive imaging methods including magnetic resonance angiography (MRA), colour Doppler ultrasound (CDU), computerized tomography angiography (CTA), PET with 18F-fluorodeoxyglucose (18F-FDG) and 18F-FDG PET/CT [^(8)] are gaining importance over DSA now-a-days. Irregular, long segment smooth tapered stenosis ranging from mild to severe and complete occlusions, collateral formation or subclavian steal phenomenon can be demonstrated 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, substantial radiation dose. It is difficult to perform in long segment stenosis. The complications DSA are eg. Vascular injury, pseudoaneurysm, catheter breakdown, coiling etc.

However, it's disadvantage is that it only shows the lumen of vessels; & information about the arterial wall is not obtained.

Classification

Angiographic classification of Takayasu's arteritis - 6 Type Vessel involvement.[^(9)]

Type I Branches from the aortic arch

Type IIa Ascending aorta, aortic arch, and its branches.

Type IIb 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 IIb and IV.

Involvement of the coronary or pulmonary arteries should be designated as C (+) or P (+) respectively.

Treatment

MEDICAL

TA is a chronic systemic inflammatory disease which undergoes multiple remissions and exacerbations. High dose systemic steroids such as Prednisolone are indicated in active phase to control disease activity. Immunosuppressive agents such as Methotrexate, Azathioprine, Mycophenolate Mofetil, Cyclophosphamide, Cyclosporin-A are also used in the medical management of TA.

Surgical

Angioplasty and Bypass surgery is to be considered only when there is no active inflammation. Symptomatic fibrotic lesions (stenosis or occlusion) can be managed by interventions such as angioplasty or stent placement. In severe cases, vascular resection and surgical placement of composite grafts are preferred. Prognosis varies from patient to patient such as rapidly progressive disease in some, where as quiscent disease in some.

V. Conclusion

TA is a chronic form of large vessel vasculitis resulting in stenotic, occlusive lesions involving arterial walls leading to a highly variable spectrum of presentation, varying stages of disease activity & response to treatment. Digital subtraction angiography has an important contribution in demonstration of arterial involvement in the form of stenosis, occlusion, dilatation as well as has appropriate therapeutic options like angiography with stenting in the inactive phase. DSA is also used for follow up for disease progression, remission and to check efficacy of interventional therapeutic modalities. Most of our patients were not fit for primary angiographic intervention (due to active stage of disease). Therefore appropriate medications like NSAIDS, steroids and in some cases immunosuppressive agents were prescribed. The patients were also informed about the waxing and waning course of the disease and the need to alter the medication dosage accordingly. They were also made aware about the side effects of such treatment like infections, adrenal suppression, cataracts, hyperglycemia, hypertension, osteoporosis and aseptic necrosis. All patients were couoncelled accordingly.

References

[1]. Rossman MG (2011) Takayasu arteritis. http//emedicine.medscape.com/article332378-overview. Accessed 27 Sep 2011
[2]. Subramanyan R, Jay J, Balakrishan KG. Natural history of aortoarteritis (Takayasu's disease). Circulation 1989; 80: 429-37.
[3]. Takayasu. A case with peculiar changes of the central retinal vessels. *Acta Societatis ophthalmologicae Japonicae*, Tokyo 1908, 12: 554.
[4]. Moriwaki R, Moda M, Yajima M et al. Clinical manifestations of Takayasu's arteritis in India and Japan – new classification of angiographic findings. Angiology 1997; 48: 369-79
[5]. Miller DV, Maleszewski JJ. The pathology of large-vessel vasculitides. Clin Exp Rheumatol. 2011;29:S92–8. [PubMed]
[6]. Nastri MV, Baptista LP, Baroni RH et-al. Gadolinium-enhanced three-dimensional MR angiography of Takayasu arteritis. Radiographics
[7]. Dähnert W. Radiology Review Manual. Lippincott Williams & Wilkins. (2011)
[8]. MavrogeniS, DimitroulasT, ChatziioannouSN, et al. The role of multimodality imaging in the evaluation of Takayasu arteritis, Semin Arthritis Rheum, 2013, vol. 42(pg. 401-12)
[9]. Gotway MB, Araoz PA, Macedo TA et-al. Imaging findings in Takayasu's arteritis. AJR Am J Roentgenol. 2005;184 (6): 1945-50

ABREVIATIONS

BA – Brachial Artery, CCA - Common Carotid Artery, DSA – Digital Subtraction Angiography, ECA – External Carotid Arrtery, ICA – Internal Carotid Artery, LUL –Left Upper Limb, LOC – Loss Of Consciousness, MCA – Middle Cerebral Artery, RUL – Right Upper Limb, TA – Takayasu Arteritis.

Dr. Debashis Dakshit "An Original Article Spectrum of Imaging Findings of Takayasu Arteritis With Digital Subtraction Angiography : A Prospective Study In A Tertiary Care Hospital."IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 17, no. 4 2018, pp 52-61.