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RESEARCH ARTICLE

CLINICAL PROFILE, ANGIOGRAPHIC FINDINGS AND OUTCOMES FOLLOWING INTERVENTION IN PATIENTS WITH TAKAYASU'S ARTERITIS.

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Abstract

Objective: To analyse the clinical presentation, angiographic findings and outcome following intervention (endovascular and open surgical) in patients with Takayasu's arteritis.

Methods: This is a retrospective observational study of 28 patients who were diagnosed with Takayasu's Arteritis and who presented to the department of Vascular Surgery, between January 2012 to January 2018. Their clinical presentation , angiographic findings, and outcome following intervention was analyzed.

Results: 28 patients were analysed during study period. Among 28 patients, 25(89%) were female and 3(11%) were male. Average age of presentation was 29.34 years. Constitutional symptoms were reported in 13 patients(46.43%) and claudication in 11 patients(32%) .The commonest finding was hypertension in (57%) followed by absent pulses in 10 patients (42%). Major organ involvement was seen as cerebrovascular accident in 1 patient(7.14%) and cardiac findings included - chest pain 2(10.71%), aortic regurgitation 4(14.29%), mitral regurgitation or valve prolapse 2(7%) and pulmonary hypertension 2(7%) .

According to the new angiographic classification,type I(32.14%) was encountered most frequently, followed by type III (25%), type IV (21.4%), type V (14%) and type IIb(7%) .The commonly involved vasculature was abdominal aorta(32%) , renal artery(25%) ,left subclavian (25%) followed by thoracic aorta(14%) and right subclavian (10%). Renal artery angioplasty was done in 5 patients and aneurysm repair was done in 1 patient with good outcome.

Conclusion:-Takayasu's Arteritis is a rare disease, with female predominance and varied clinical presentation. Angiography findings and clinical symptoms are integrated to decide management. Angiographic evaluation and percutaneous transluminal angioplasty with stenting is useful in selected cases; while majority of the patients are managed medically.

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Introduction:-

Takayasus arteritis is a chronic inflammatory disorder of the aorta and its branches which can present with varied clinical manifestations making diagnosis challenging(1,2). Progressive inflammation can lead to vessel wall thickening, stenosis, aneuryms which present clinically as features of end organ ischemia(3). Angiography is the gold standard for diagnosis, aided by inflammatory markers(4).The New Angiographic Classification classifies it into 5 types. Medical management with steroids or methotrexate is the mainstay of treatment. Surgical intervention which includes angioplasty and aneurysm repair is indicated in severe extremity claudication ,hypertension with critical renal artery stenosis, severe cardiac or cerebral involvement (3). Treatment is aimed at controlling disease activity and to maintain vascular competence(3).

Materials And Methods:-

This is a single centre retrospective observational study which analysed the clinical and angiographic profile of 28 patients with Takayasu's arteritis between January 2012- Jan 2018. All patients fulfilled the proposed criteria(as given below) of the American college of Rheumatology for diagnosis of Takayasu's arteritis. Details on clinical symptoms, haematological parameters and imaging findings were obtained from the hospital records, and follow up of patients in OPD.

Modified diagnostic criteria for Takayasu's arteritis:-

Three Major Criteria:-

Left mid subclavian artery lesion
Right mid subclavian atery lesion
Characteristic signs and symptoms for atleast 1 month:
Limb claudication, pulselessnes or pulse difference in the limbs, BP difference> 10, fever, neck pain, transient amaurosis, blurred vision, syncope, dyspnea, palpitation.

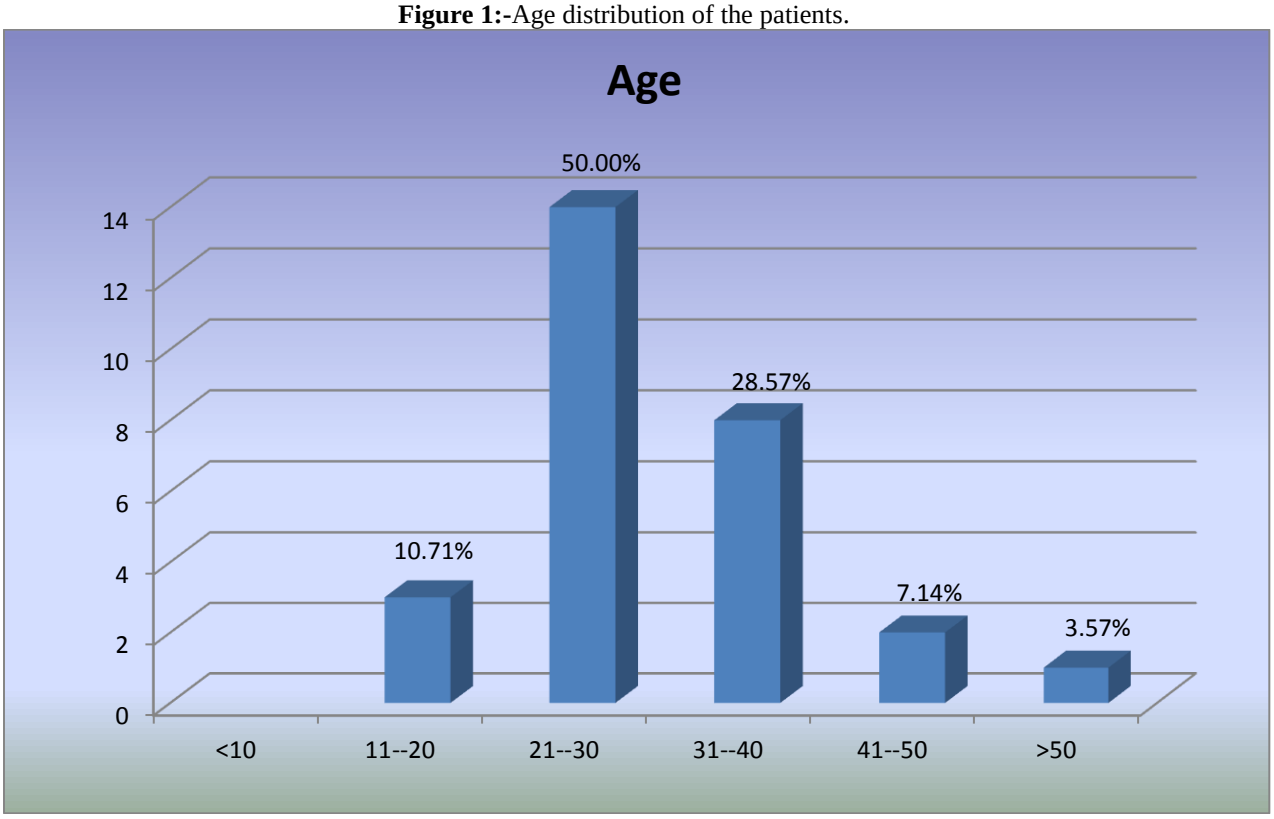
Ten Minor Criteria:-

Elevated ESR
Carotodynia
Hypertension
Aortic regurgitation
Pulmonary artery disease
Left mid common carotid disease
Distal brachiocephalic trunk lesion
Descending aortic lesion.
Abdominal aortic lesion
Coronary artery lesion

Results:-

28 patients of Takayasu's arteritis were evaluated. 89% of patients were female (25-Female, 3-Male, F/M: 8.3/1). The mean age was 29.34 years at the time of diagnosis. The age wise distribution is given in figure 1.

Figure 1:-Age distribution of the patients.



The clinical presentation of the patients is summarised in table 1. Constitutional symptoms were reported in 13 patients(46.43%) and claudication in 11 patients(32%). Hypertension in 16 patients(57.14)%. Difference in blood pressure was seen in12 patients (42.86%), and absent pulses in 10 patients (42%). Major organ involvement was seen as cerebrovascular accident in 1 patient(7.14%) and cardiac findings included- chest pain 2(10.71%), aortic regurgitation 4(14.29%), mitral regurgitation or valve prolapse 2(7%) and pulmonary hypertension 2(7%) .

Table 1:- Demographic details and clinical presentation

Characteristic	n (%)
Female	25(89%)
Male	3(11%)
Average age at onset (years)	29.34
Constitutional symptoms	13(46.43%)
Fever	8 (28.57%)
Headache	12 (42.86%)
Weight loss	2(7.14%)
Myalgia	6 (21.43%)
Arthralgia	0
Chest pain	3(10.7%)
Abdominal pain	2(7.14%)
Absent pulses	
Upper limbs	9(32.14%)
Lower limbs	3(10.71%)
BP difference	12(42.86%)
Carotodynia	1(3.57%)
Hypertension	16(57.14)
Claudicaition	
Upper limbs	9(32.14%)

Lower limbs	3(10.71%)
Gangrene	0
Ocular complaints	1(3.57%)
Stroke	1(3.57%)
Cardiac	
Aortic regurgitation	4(14.29%)
Mitral regurgitation	1(3.57%)
Mitral valve prolapse	1(3.57%)
Myocarditis	0
Pulmonary hypertension	2(7.14%)
Elevated ESR	16(57.14%)

Figure 2:-Presentation pattern.

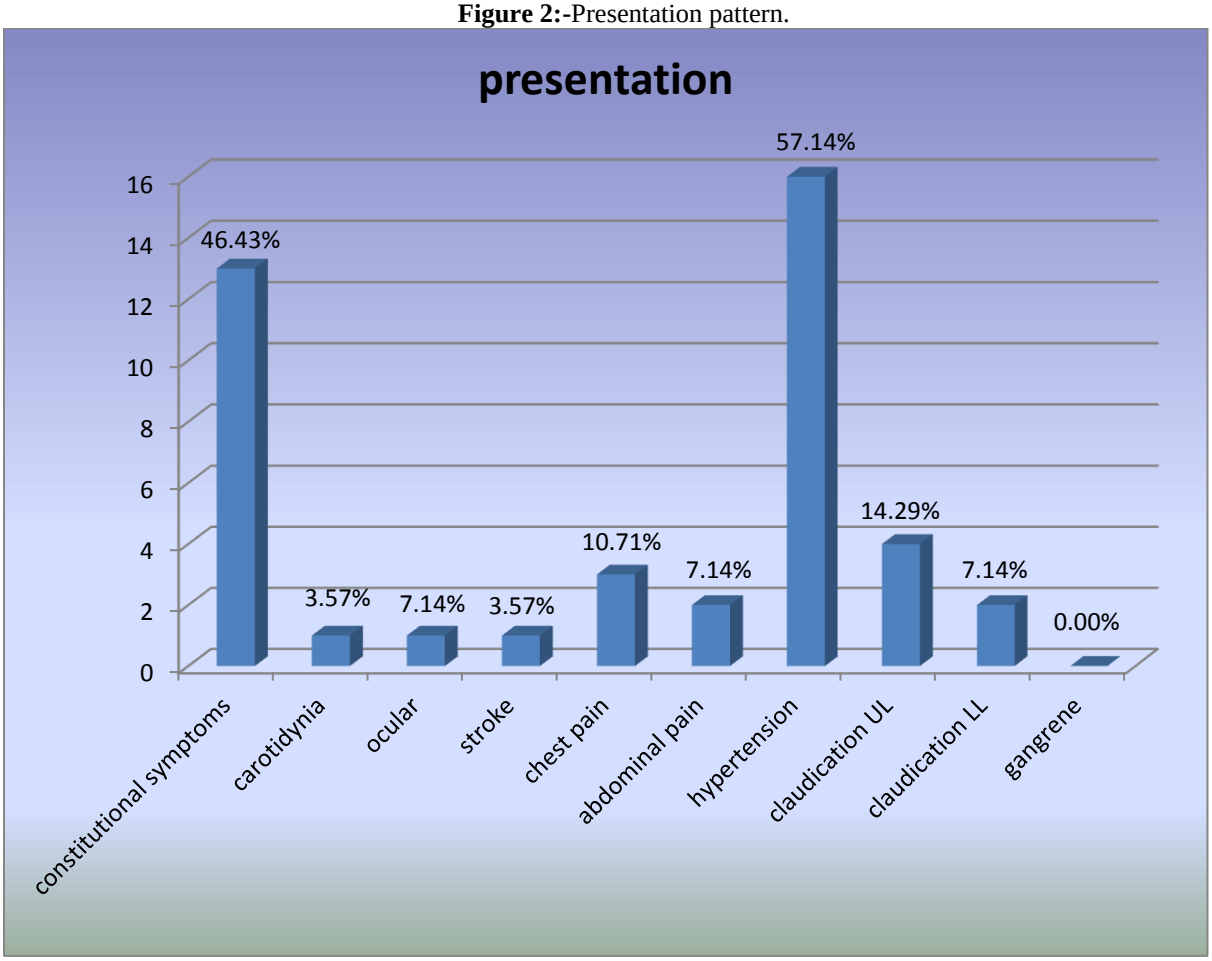


Figure 3:-constitutional symptoms

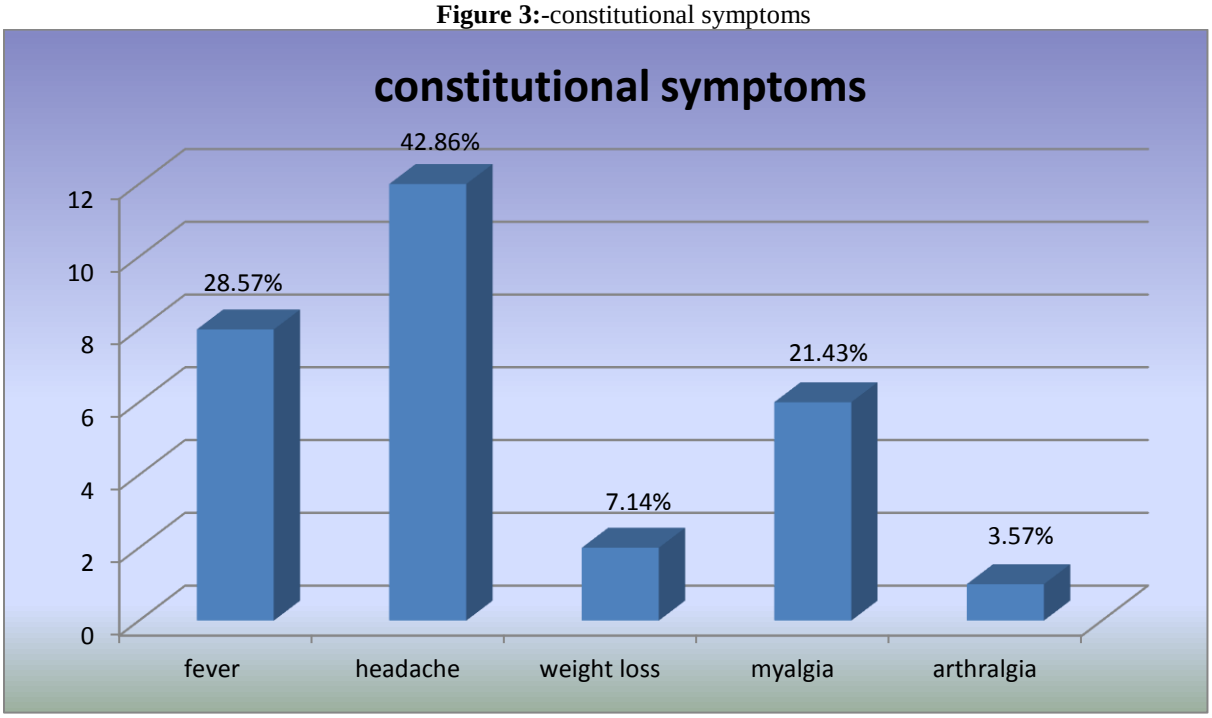
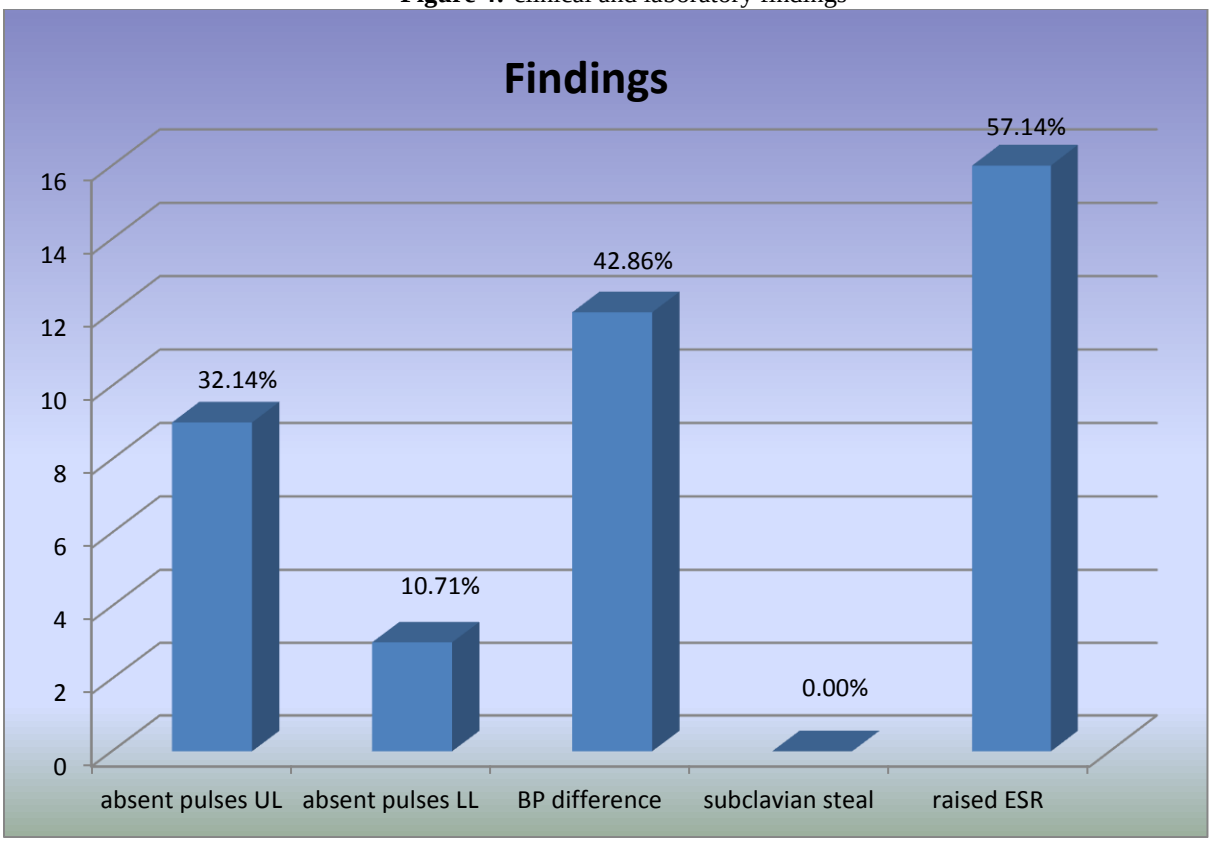


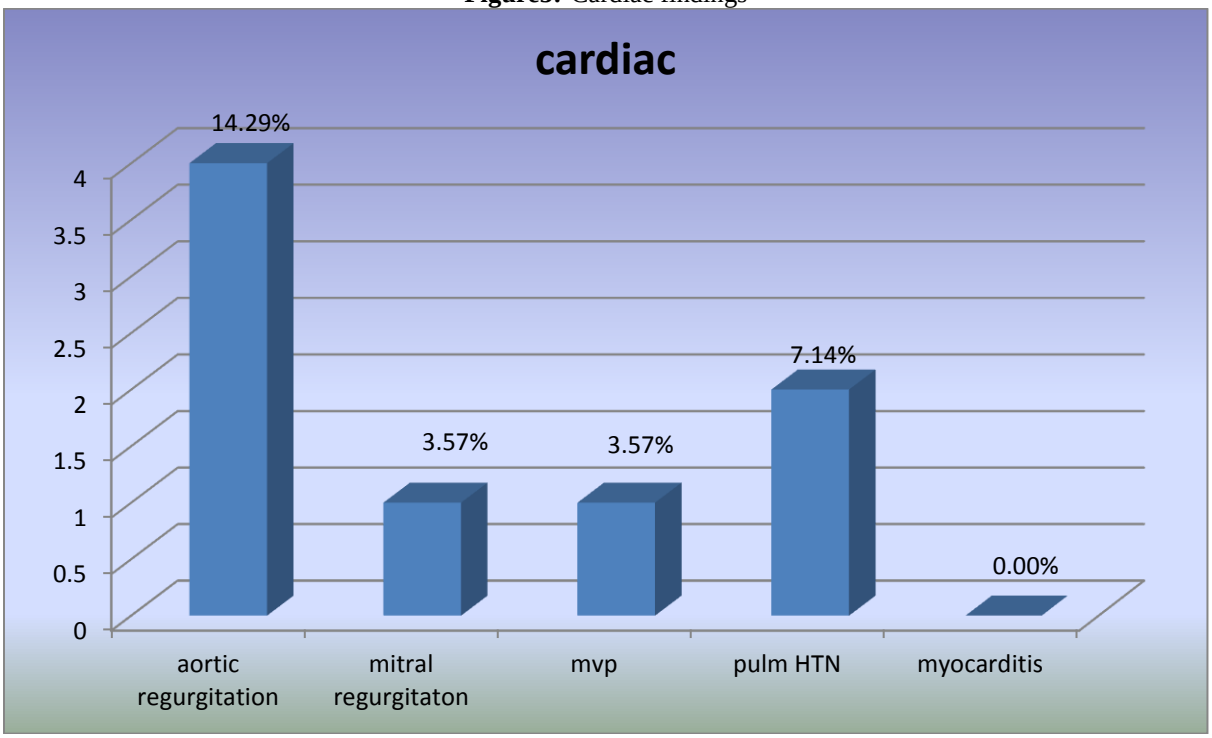
Figure 4:-clinical and laboratory findings



Patients mostly presented with constitutional symptoms (46%) of which headache was seen in 12 patients (42%). Hypertension was seen in 16 patients(57%). Absent pulses in the upper limb was seen in 9 patients(32%), though

claudication pain was not a usual complaint among the patients. 16 patients(57%) patients presented with elevated ESR.

Figure5:-Cardiac findings



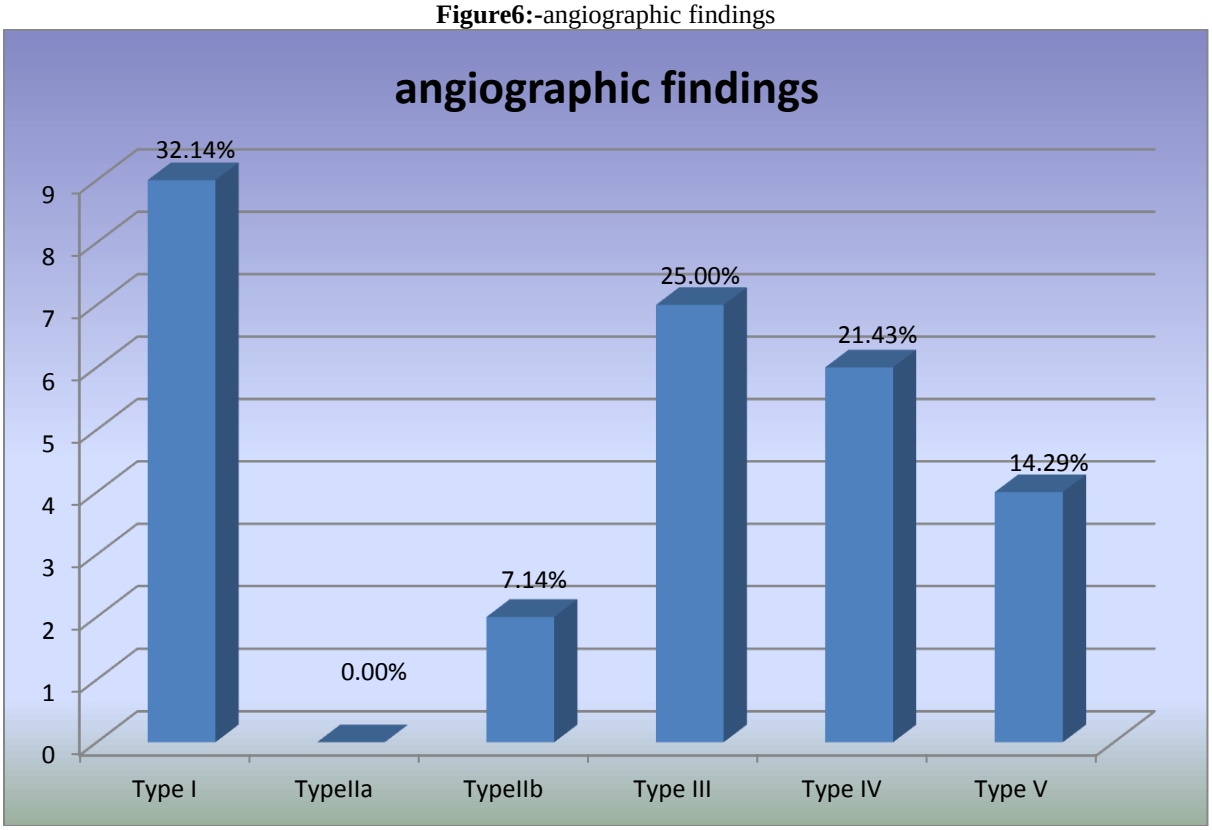
Echocardiogram was done as a routine in all our patients(n=28). Aortic regurgitation was seen in 4 patients (14%). Mitral regurgitation and mitral valve prolapse was seen in 1 patient each (3%). Pulmonary hypertention was seen in 2 patient (7%) . Coronary angiogram was done only for one patient who was planned for a thoraco abdominal aneurysm repair. Coronary angiogram showed right dominant system with normal epicardial coronaries.

According to the new angiographic classification,angiographic type I (32.14%) was encountered most frequently, followed by type III (25%), type IV (21.4%), type V (14%) and type IIb(7%).

Table 2:-Angiographic findings

Type I	9(32.14%)
Type IIa	0
Type IIb	2 (7.14%)
Type III	7 (25.00%)
Type IV	6 (21.43%)
Type V	4 (14.29%)

Figure6:-angiographic findings



The common vessels involved were abdominal aorta(32%) , renal artery(25%) and left subclavian (25%) followed by thoracic aorta(14%) and right subclavian (10%).

Table 3:-Vessel involvement

Right subclavian	3(10%)
Left subclavian	7(25%)
Carotid artery	2 (7%)
Thoracic aorta	4 (14%)
Abdominal aorta	9 (32%)
Renal artery	7 (25%)
Mesenteric vessels	1 (3%)
Common iliac , femoral artery	2 (7%)

Renal artery plasty was done in 5 patients and aneurysm repair was done in 1 patients with good clinical reponse during the follow up period. Anti hypertensives could be discontinued in 2 patients and remaining 3 patients required only single antihypertensive. Redo angioplasty was done for 1 patient. Thoracoabdominal aneurysm repair with mesenteric vessel reimplantation was done for 1 patient. All other patients were treated medically with steroids or methotrexate.

Discussion:-

Takayasu's arteritis is a rare inflammatory disorder affecting aorta and its branches. Chronic inflammation of the vessel wall leads to vascular incompetence in the affected organ or body part(5).Females are affected eight times more than men, which is seen in our study also(5). The vascular lesions result from chronic inflammation leading to fibrosis of all three layers leading to patchy narrowing. Rapid progression can lead to vessel wall destruction with aneurysm formation. (3). Microscopically, the vessel wall is infiltrated with lymphocytes and giant cells with subsequent thickening of the intima by smooth muscle cells and fibroblasts(3).

The clinical manifestation of the condition is very varied from asymptomatic disease picked up as absent pulses and hypertension or can present as severe ischemic complications (4). The clinical presentation in our study has been found to be similar to other observational studies done by Setty et al and Li et al; commonly encountered are constitutional symptoms, Hypertension, unequal pulses, BP difference and limb claudication. Rarer symptoms include limb gangrene, carotodynia and major organ involvement causing cerebrovascular accidents, bowel ischemia and heart valve impairment and coronary syndromes(4,6). The 1990 ACR diagnostic criteria used for the diagnosis of TA, given in the table below(7):

Table 4:-1990 ACR Criteris for diagnosis of Takayasu's Arteritis

1.	Age of onset ≤40
2.	Limb claudication
3.	Diminished brachial pulse
4.	Difference of >10mm Hg systolic
5.	Bruit over the subclavian artery or aorta
6.	Narrowing or obstruction in the aorta, its main branches or large vessels in proximal part upper or lower extremities which have been confirmed angiographically.
	For diagnosis ≥ 3 criteria should be present

Angiogram is the gold standard for diagnosis for a suspected case of Takayasu's arteritis. The angiographic findings are classified as per table 5

Table 5:-The New Angiographic Classification of Takaysu's Arteritis

Type	Vessel Involvement
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 IIB and IV

Studies done in Japanese patients show a predominance of ascending aorta and aortic arch (Type 1, IIA) whereas Indian patients had a higher frequency of abdominal aorta and renal artery (type IV) involvement(8,9). Similar finding in Indian patients was reported by Sharma et al with renal artery involvement in 76% of the patients and type 1 only in 10% of the patients. Our study shows a higher frequency of Type 1(32.14%), followed by Type III(25%) and Type IV(21.4%). The commonest vessel involved was abdominal aorta followed by renal artery and left subclavian. Our findings are similar to study by Setty et al done in a South Indian centre with maximum Type I involvement(40%), followed by Type III(30%).

The level of evidence for management of TA is low as there are no placebo controlled randomised controlled trial. In a systematic review by Keser et al, most of the patients were treated with medical management with high dose steroids(1mg/kg) followed by immunosuppressive agents such as Methotrexate, Azathioprine and Mycophenolate mofetil for steroid refractory cases. Antiplatelets can be used to reduce ischemic complications (10). Surgical intervention include balloon angioplasty or stent placement for short segment, critical stenosis and surgical bypass for long segment stenosis with extensive periarterial fibrosis. Thoracic aneurysm repair is also advised if technically possible. Intervention is avoided in the active phase of the disease(3,10). In our study only 6 out of 28 patients underwent surgical intervention- 5 renal artery stenting and 1 aneurysm repair. All other patients were treated medically.

Conclusion:-

Takayasu's arteritis is a rare clinical entity which requires high clinical suspicion and early diagnosis to prevent chronic or catastrophic complications. The findings of the imaging modality, patient presentation and lab tests should be integrated to guide management and prognosticate.

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