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Review Article

CLINICAL AND IMAGING PROFILE OF PATIENTS WITH TAKAYASU ARTERITIS – A REVIEW OF CASE RECORDS

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A B S T R A C T

Background: Takayasu arteritis is a rare chronic inflammatory granulomatous vasculitis involving the aorta and its major branches which leads to intimal fibrosis and narrowing of the vessels. Different patterns of involvement of the arterial system in patients have been reported from various parts of the world. Clinical presentations may vary according to the vessels involved.
Methods: Review of the case records of the patients fulfilling the American College of Rheumatology classification criteria for Takayasu arteritis that attended the Rheumatology clinic during the period from June 2013 to October 2019 was done and the data were analysed .We have done the literature review and compared our data with the other studies done in various parts of the world.
Results: There were 64 patients with a male, female ratio of 1:4 .Mean age at the time of diagnosis was 29.25±9.9 yrs. There were 4 children. Claudication pain was seen in 50% of patients. Absent or feeble pulse was noted in 75% of the patients. Acute phase reactants were elevated in 60% of patients. Type I angiographic type was seen in 27% followed by Type V in 25% of patients.
Conclusion: Commonest clinical presentation was claudication pain. Cardiac involvement was found to be the commonest systemic complication with valvular regurgitation being the commonest lesion. Angiographic type I was found to be the commonest type.

INTRODUCTION

Takayasu's arteritis (TA) is arare systemic large vessel vasculitis with granulomatous inflammation in the adventitia and media of the aorta and its major branches (1). In Germany the estimated annual incidence of TA is 0.4–1.0 per million people (2), and in south east Norway it is 2 per million (3).In the UK primary care cohort, the prevalence is 4.7 per million (4), but it is more common in southeast Asia, India, Japan, and Mexico [5]. The prevalence of TA in Japan was reported to be 40 per million [6]. In the United Statesthe incidence of TA was 2.6 cases per million population annually (7). Various types of Clinical presentations have been reported from different parts of the world.

MATERIALS AND METHODS

We have done the review of the case records of the patients fulfilling the American College of Rheumatology classification criteria for Takayasu Arteritis that attended the Rheumatology clinic in a tertiary care teaching hospital in South India during the period from June 2013 to October 2019.CT angiogram of Aorta, MR aortography, MR Angiography of cerebral vessels and arterial Doppler studies were the imaging modalities used. Demographic profile, clinical features, laboratory parameters and imaging studies were analysed and literature review was done. We have compared our data with the other data reported by the studies done in various parts of the world. Institutional Ethical committee approval waived.

RESULTS

Out of 64 patients there were 12 (19%) males and 52 (81%) females with the ratio of 1:4. There were 4 children. Mean age of the patients at the time of hospital visit was 32 ± 10.7 yrs. Mean age at the time of diagnosis was 29.25±9.9 years. Mean duration of the disease related symptom at the time of diagnosis was 32.7 months. The presenting symptoms are tabulated (Table 1). Clinical examination findings included absent or feeble pulse in 48 (75%), Carotid bruit in 20 (31%) and renal bruit in 12 (19%) patients. Carotidynia was noted in one patient. Hypertension was noted in 21 (33%) patients. Acute pulmonary edema was noted in 2 patients. Congestive cardiac failure was noted in 3 patients. Laboratory studies

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showed elevated inflammatory markers in 33 (60%) out of 55 patients for whom the inflammatory markers were done Seven patients (13%) had raised Erythrocyte Sedimentation rate and 14 (25%) patients had elevated C - reactive protein. Both ESR and CRP were elevated in 12 (22%) patients. Anemia was noted in 5 patients. Systemic complications were noted and presented in

Table 1 Presenting symptoms of patients with Takayasu arteritis in our study

Clinical feature	No (%)	Clinical features	No (%)
Vascular Symptoms			
Claudication	32 (50)	**Others**	
Neurological Symptoms		Fever	1 (1.6)
		Fatigue	1 (1.6)
Head ache	7 (11)	Hemoptysis	1 (1.6)
Seizures	3 (5)	**Asymptomatic**	2 (3)
Syncope	1 (1.6)		
Weakness of the limbs	5 (8)		
	1 (1.6)		
Vertigo	1 (1.6)		
Diplopia	1(1.6)		
Cardiac Symptoms			
Chest Pain	7(11)		
Dyspnea	4 (6)		
Palpitation			
Visual Symptoms			
Impaired Vision	3(5)		
Visual Loss	4(6)		
Musculo skeletal Symptoms			
Joint Pain	6(9)		

Table 2 Prevalence of systemic complications in our patients with Takayasu arteritis

Systemic Complications	No (%)	Systemic Complications	No (%)
Cardiac Complications			
Aortic Regurgitation	7 (11)	**Cerebro vascular accidents**	
Mitral Regurgitation	7 (11)	Ischemic Infarcts	7 (11)
Coronary artery disease	4 (6)	SAH	1 (1.6)
Dilated Cardiomyopathy	7 (11)	TIA	1 (1.6)
Pulmonary arterial hypertension	2 (3)	PRES	1 (1.6)
Left ventricular Hypertrophy	4 (6)	**Obstetric complications**	3 (5)
		IUD	2 (3)
Ocular Complications		Abortions	3 (5)
Hypertensive retinopathy	13 (20)	PIH	1 (1.6)
CRAO	2 (3)	Primary Infertility	
Retinal Vasculitis	2 (1.6)	**Others**	2 (3)
Neovascularisation	2 (3)	Renal failure	2 (3)
Ocular ischemic syndrome	1 (1.6)	Serositis	1 (1.6)
Venous dilatation with arteriolar attenuation	1 (1.6)	Renal infarct	1 (1.6)
Retinal haemorrhage	1 (1.6)	Splenic infarct	1 (1.6)
Scleritis	3 (5)	Acute Psychosis	

CRAO-Central retinal artery occlusion, SAH-Sub arachnoid haemorrhage, TIA-Transient ischemic attack, PRES-Posterior revesible encephalopathy syndrome, IUD-Intra uterine death, PIH-Pregnancy induced hypertension

Table 2. Angiographic types were analysed and presented in figure 1. Frequency of involvement of various vessels are presented in Table 3. Aneurysmal dilatation of aortic root and other parts of the aorta were noted in 3 (5%) and 7 (11%) patients respectively.

Table 3 Frequency of involvement of major vessels in our study population

Type of the vessel involved	No (%)		No (%)
Left CCA		MCA branches	4(6)
Right CCA	15(23)	ACA	2(3)
Both CCA	4 (6)	Left vertebral Artery	5(8)
Left SCA	17(27)	Right vertebral Artery	1(1.6)
Right SCA	19(30)	Both vertebral Arteries	1(1.6)
Both SCA	5(8)	Right renal artery	5(8)
Right ICA	8(13)	Left renal artery	9(14)
Left ICA	4(6)	Both renal arteries	7(11)
Both ICA	5(8)	SMA	7(11)
Left ECA	4(6)	Coeliac Trunk	7(11)
Right ECA	0	Pulmonary artery	2(3)
Both ECA	2(3)		
Innominate Artery	12(18)		

CCA-Common carotid artery, SCA- Subclavian artery, ICA-Internal carotid artery, ECA-External carotid artery, MCA-Middle cerebral artery, ACA-Anterior cerebral artery, SMA-Superior mesenteric artery

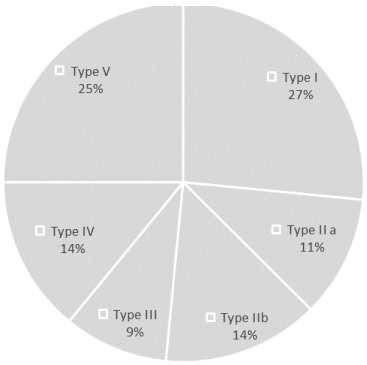

Figure 1 Prevalence of angiographic types of Takayasu arteritis in our Study

DISCUSSION

Takayasu arteritis is a chronic inflammatory granulomatous vasculitis involving the aorta and its major branches which leads to intimal fibrosis and narrowing of the vessels. Aneurysmal dilatation may also develop in the affected vessels. There are six angiographic types classified according

to the type of vessel involvement. Clinical presentations may vary according to the vessels involved. Most of the patients are diagnosed at a later stage when the disease had progressed to cause stenosis of the affected vessels. Females are more commonly affected. In our study 81% of patients were females which is similar to a study that was done by Setty *et al* (8). The female, male ratio was similar to a study done in China (9). Haiying Wu *et al* reported the onset age ranging from 2 to 68 years with the mean age of 28.9 ± 12.0 and the largest proportion of their patients was in the age group between 20 and 40 years (10).We have found highest number of patients in the age group between 30 and 40 years in our study.Takayasu arteritis may be asymptomatic sometimes, may have subtle symptoms or present with constitutional symptoms alone during the early inflammatory phase. Inflammatory phase is followed by Pulseless phase when the patients present with vascular symptoms and signs due to the narrowing of the vessels. In our study vascular claudication was the commonest symptom and was noted in 50% of the patients. Arm claudication or numbness was found in 37% of patients in a study done by Islam *et al* (11). Dizziness was reported to be the commonest initial presenting symptom (37.8%), followed by vascular claudication (28.4%) in a study done by Liuri *et al* (12). Hypertension was noted in 33 % of our patients and 57 % of them hadrenal artery stenosis. Mammery A *et al* have reported the prevalence of hypertension as 45.8% in their study and found renal artery stenosis in 54% of them (13). All our patients with obstetric complications were found to have hypertension. Arterial bruit was noted in 50% of our patients. Cardiac involvement was the commonest complication which was noted in 48 % of the patients. Valvular regurgitation was the commonest lesion among them (22%). Prevalence of valvular regurgitation was reported to be35% in a study done by Zhang Y *et al*, which included 1069 patients. Among them aortic regurgitation was found in 69.7 %, mitral regurgitation in 39.1% of patients, tricuspid regurgitation in 34.6% and pulmonary regurgitation in 11.8%. Valve stenosis was reported to be rare (14). In our study aortic and mitral regurgitation were seen in equal number of patients. Carotidynia was the least observed clinical feature in our study. It was observed in 4.4% of patients in a study done by Lirui *et al* (12). Ocular complications were seen in 38% of our patients. Ocular manifestations in TA are due to i) ischemic ocular changes secondary to arteritis and ii) retinopathy secondary to hypertension. Chun YS *et al* have found hypertensive retinopathy in 48 (30.8%) eyes and Takayasu retinopathy in 21 (13.5%) eyes in a study involving 156 eyes in 78 patients. Blindness may occur in TA due to ischemia, vitreous haemorrhage, retinal detachment, or optic atrophy (15). Takayasu retinopathy was seen in 15%, ocular ischemic syndrome in 7%, and hypertensive retinopathy in 16% of patients in a study done by Peter J *et al* (16).In our study 14 % had Takayasu retinopathy and 20% had hypertensive retinopathy. Central nervous system (CNS) complications were seen in 17% of our patients. Cerebral Ischemic infarct was the commonest CNS lesion noted. Cerebrovascular accidents may be the first presenting manifestation in TA(17). Four of our patients presented with cerebral ischemic infarct at the time of diagnosis. Angiographic type I was the commonest type noted in our study which was seen in 26% followed by Type V in 25 % of patients. Left Subclavian artery was the commonest vessel involved in our patients. Singh AP *et al* havereported Type V as the commonest type (37%) and the left Subclavian artery as the commonest vessel involved in their study (18). Clinical examination of patients attending the general medical clinic should include examination of peripheral pulses for absence, inequality, carotidynia and bruit. Early diagnosis and initiation of appropriate treatment will prevent most of the complications due to Takayasu arteritis.

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