TAKAYASU'S DISEASE:
ANALYSIS OF CLINICAL AND ANGIOGRAPHIC FEATURES

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Analysis of 20 cases of Takamasa's Disease is presented with a view to clinical manifestations and investigations including angiographic findings. There were 15 females and 5 males with a mean age of 21 ±7.1 years (Range 15-33 years), 90% cases presented with pulselessness while lesion of subclavian arteries was present most frequently (90%). The findings are in conformity with other studies.

INTRODUCTION

Takayasu's disease is a chronic inflammatory disease of unknown origin with a worldwide distribution and

is prevalent in females[12]. It causes luminal narrowing and/or dilation of aorta and or its main branches. Pulmonary arteries may also be involved in about
50% of cases [3,4,5]

The disease has four main complications i.e. ischemic retinopathy, hypertension, aortic regurgitation and aneurysm formation [6,7,8]. Coronary arteries are rarely affected. The mortality is mainly dependent upon the severity of cardiovascular complications which vary from patient to patient.

Regarding nomenclatures, confusion has been caused by various authors who gave different names to the same disease process. Because of difficulty in applying the various diagnostic criteria, it has been called pulseless disease, aortic arch syndrome, idiopathic arteritis, reverse coarctation, Mortelli's Syndrome etc. It has eventually been universally accepted to be called as Takayasu's disease (Thrombo-aorto-arteriopathy) [4,6,9,10]

Despite extensive work on clinical and pathological process of the disease, its exact etiology has still not been fully elucidated. Initially the disease process was thought to be limited to the aortic arch and its main branches but subsequent studies have also shown the involvement of the descending aorta and its branches [4,5,11,12].

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MATERIALS AND METHODS

The diagnosis of Takayasu's disease was established by clinical and angiographic study in all the cases. Arterial biopsy or autopsy was not done on any patient. Pulmonary arteriography was available on two patients only. All patients had detailed clinical history and thorough physical examination with routine investigations like complete blood count, ESR, urea, electrolytes, ECG and chest X-ray, echo-cardiograms were available on only 12 patients, ultrasound with Doppler was carried out in 5 cases. Functional disability was classified according to New York Heart Association (NYHA) classification. Cardiomegaly on chest X-ray was defined as cardio- thoracic ratio of more than 50%. Aortic aneurysm was defined if the vessel diameter was double the size of normal. Left ventricular hypertrophy on electrocardiogram was defined when sum of "S" and "R" waves amplitude in V1 and V6 was more than 35 mm.

Aortic and Mitral Regurgitation were graded on colour Doppler echocardiography. Retrograde aortography was performed percutaneously through right femoral artery with Seldinger's technique. For thoracic aortography a pigtail catheter tip was placed at the aortic root and 30 ml of radio contrast was injected at rate of 15 ml/sec. For thoraco-abdominal aortography the catheter tip was placed in the thoracoabdominal aorta at the level of 11th thoracic vertebra and 20 ml of dye was injected at rate of 10 ml/sec. Selective catheterization of common carotid and subclavian was done with multipurpose catheter according to clinico-cardiological circumstances. Coronary angiography was not done on any patient. Pulmonary arteriography was performed on two patients.

The main sign of aorto-arteritis was pulselessness described as absent or diminished pulse, difference in pulses in upper limbs or blood pressure difference (> 10 mm of Hg) in two limbs

RESULTS

20 in-patients were studied in the Department of Cardiology, Postgraduate Medical Institute/Lady Reading Hospital, Peshawar over a period of 11 years. There were 15 females and 5 males with female to male ratio of 3:1. The mean age was 21. ±7.1 (Rage 15-35 years).

The clinical features on admission are shown in Table-1.

TABLE-1: CLINICAL FEATURES ON ADMISSION

Clinical Features	No of Pts	%age
1. Constitutional symptoms of weakness, aches, pains etc.	15	75
2. Cardiovascular symptoms/ signs		
i) Functional disability		
a. Mild NYU A class I or II.	18	90
h. Severe NYHA class III or IV.	2	10
li) Palpitation.	7	35
iii) Chest pain-cause?	3	15
iv) Intermittent claudication.	5	25
v) Vascular bruit.	12	60
vi) Carotid artery tenderness.	3	15
vii) Pulse deficit.	18	90
viii) 1 high BP 140/90 mm of Hg	9	45
ix) Aortic Regurgitation.	2	10
x) Mitral Regurgitation & heart failure.	1	5
3. Neurological symptoms and signs.		
i) Headaches.	4	20
ii) 1 lemiplegias. TIAs.	3	15
iii) Visual disturbances and fundal changes.	6	30

The laboratory, radiological and ECO findings arc given in Table-2.

TABLE-2: INV ESTIGATIVE WORKUP

Investigations	No of Pts	%age
1. Laboratory:		
i) Anaemia (Hh 1 Ogm/dl).	7	35
ii) KSR 40mm 1st hour.	5	25
iii) Blood urea 60 mg%.	2	10
2. Radiological Chest X-ray Findings:		
i) Aortic knuckle prominence.	10	50
ii) Aortic calcification.	3	15
iii) Thoracic aortic wall irregularity.	2	10
iv) Rib notching.	4	20
v) CTR 50° «.	7	35
3. Electrocardiographic Findings.		
i) Left ventricular hypertrophy.	6	30
ii) I,eft bundle branch block.	2	10
iii) Left atrial enlargement	1	5

Carotid Doppler ultrasound was available on five patients, three showed diffuse narrowed segment of carotid arteries with high turbulent flow velocities.

Angiographic findings are given in Table-3 which show the subclavian arteries to be most commonly involved. No patient had ileal or ileo-femoral involvement. Collateral circulation was demonstrated in 80% (1 Ceases). Aortic coarctation was seen in 6 patients (30%). Two patients (10%) developed diminished pulsations in right femoral artery after the aortography and complained of intermittent claudication but there was no procedure or operative death.

Angiographic arterial lesion distributions are shown in Table-3.

Angiographic Arterial Lesions Site.	No of Pts.	%age
1. Ascending aorta.	3	15
2. Descending thoracic aorta.	7	35
3. Abdominal aorta.	8	40
4. Subclavian arteries.	18	90
5. Carotid arteries.	11	55
6. Brachio-cephalic trunk.	9	45
7. Renal arteries.	3	15

DISCUSSION

The aim of our study was to document the clinical and angiographic features of the disease in our population. The findings are quite consistent with the results of other studies. Sex distribution in our study i.e. female to male ratio is different than that of internationally presented prevalence [2,4,1]

Inflammatory signs of active arteritis are present in early stage which was missing in our patients as majority of these presented late during the disease when fibrosis and narrowing of the vessels have occurred. This was evident by normal ESR and nontender pulses in the majority of our patients. Major clinical features of hypertension, aortic regurgitation, aneurysm formation and retinopathy were found in only small number of patients, so their severity could not be graded.

CONCLUSION

The clinical experience of retrospective study of 20 patients with Takayasu's disease is presented. The disease predominantly affected the young females. Commonest presentation was pulselessness while subclavian arteries were the most affected site on arteriography.

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