CLINICAL PROFILE OF TAKAYASU'S ARTERITIS

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ABSTRACT

Thirty consecutive cases of Takayasu's arteritis (TA) admitted over a period of four years in a large referral hospital were analysed. The extent of aorta and its branches involvement was assessed by a Duplex Doppler study and aortography. The mean age of patients was 24.7 years with the male to female ratio of 1:1.2. The vascular involvement was, Type I (10%), Type II (40%) and Type III (50%). Renal artery (76.6%), Coeliac and superior mesenteric arteries (46.6%) and right common carotid artery (30%) were the commonly involved branches of aorta. Systemic features were seen in 25 per cent cases. Hypertension (83.3%), unequal pulses (80%) and arterial bruit (66.6%) were common clinical features. Stroke (20%), cardiac failure (10%) and renal failure (10%), were the complications seen. Hypertensive retinopathy was common, but Takayasu's retinopathy was not seen.

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Introduction

Takayasu's arteritis (TA) is a non-specific inflammatory vasculitis of aorta and its branches. It causes steno-occlusive lesions or aneurysmal dilatation of involved vessels, resulting in ischaemia of various organs. The clinical features, depend on the type of aortic involvement and the organs hypoperfused. The prevalence of the disease is more in Asian countries, however incidence of TA is thought to be underestimated in the west [1]. Clinical features described in patients from Asian countries differ than those in the western countries. Even in Asia, differences in the clinical manifestations and angiographic findings have been described [2,3]. Males are more often affected than females in India as compared to Japan and the western countries. Severe and prolonged inflammatory features, absent pulses and aortic regurgitation are more frequent in Japanese patients as compared to Indians, whereas hypertension is more frequent in Indians. Aortic arch and its branches are more often involved in Japanese patients and involvement of abdominal aorta and its branches is more common in Indian patients. Investigative techniques like real-time Doppler ultrasound has helped in clinical study of these patients. This study analyses retrospectively the clinical profile of patients of TA admitted in a referral service hospital.

Material and Methods

Thirty consecutive patients of TA admitted over a period of four years from July 1993 to June 1997 in our hospital were ana-

lysed. American Rheumatology Association criteria of 1990 was used for diagnosis [4]. Detailed history, clinical examination including peripheral pulses and blood pressure in all four limbs and fundus examination were evaluated. Haemogram, ESR, ECG, chest skiagram were done in all cases. Blood for ANF was studied in nine female patients. Aortography and Doppler sonography was carried out in all cases. Aortic involvement was classified into different types according to the location of lesions as revealed in the aortogram [5]. Type I involvement consisted of patients with lesion restricted to ascending aorta, arch of aorta or its branches; Type II included patients with extensive lesions in thoraco-abdominal aorta or its branches and type III had patients with combined features of type I and II. Type IV, includes pulmonary artery involvement.

Results

The mean age of the patients at presentation was 24.7 years. The youngest and oldest patients were aged 6 years and 45 years respectively. The male to female ratio was 1:1.2. The main clinical features and complications are summarised in Table 1. Tenderness over involved arteries was not seen in any case. Two patients each had upper limb and lower limb claudication and one patient had abdominal angina. Visual blurring was reported by two patients. Jaw claudication was not reported by any patient. Hypertensive retinopathy was seen in twenty patients but Takayasu's retinopathy was not seen in any case. ESR more than 20 mm in first hour was seen in 16 patients (54%). Only three patients out of seven with systemic features had raised ESR. ANF was negative in all cases. On angiography, the vascular involvement was, Type I in 3 patients (10%), Type II in 12 patients (40%) and Type III in 15 patients (50%). Pulmonary angiography was done in only one case and it showed normal pulmonary arteries. The branches of aorta which were involved are depicted in Table 2.

Discussion

TA is generally considered to be more common in young females. The male to female ratio is reported to

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TABLE 1
Clinical features and complications of patients of Takayasu's arteritis

Clinical features	No of patients	Percentage
Hypertension	25	83.3
Unequal pulses in limbs	24	80
Abdominal bruit	12	40
Absent radial pulse	7	23.3
Systemic features*	7	23.3
Sub-clavian bruit	5	16.6
Femoral bruit	3	10
Complications		
Stroke with hemiparesis	6	20
Quadriplegia**	1	3.3
Hypertensive encephalopathy	1	3.3
Aortic regurgitation	4	13.3
Cardiac failure	3	10
Chronic renal failure	3	10

*Fever, malaise, arthralgia, weight loss and skin rash;; ** Non-compressive ischaemic cervical myelopathy

TABLE 2
Branches of aorta involved in Takayasu's arteritis

Branch	No of patients	Percentage
Renal arteries	23	76.6
Coeliac and superior mesenteric artery	14	46.6
Right common carotid artery	9	30
Left subclavian artery	5	16.6
Left subclavian artery	7	23.3
Right subclavian artery	4	13.3
Iliac	4	13.3
Femoral	6	20

be 1:8 in western countries and 1:3 in Japan [2,6]. The present study shows a much higher incidence in males. This could be due to higher numbers of males reporting to our hospital. However, similar findings have been reported by other workers from India [2,7]. The mean age at presentation was 24.7 years in this study. Type I lesion was the least common in this study. Type II and III angiographic involvement in our patients was higher as compared to patients in Japan and the west. Similar finding has been reported from other South Asian countries [8]. Disease in Japanese patients usually starts in ascending aorta and arch of aorta and descends to thoraco-abdominal aorta, whereas abdominal aorta is initially involved more commonly in Indian patients and then ascends up to involve the thoracic aorta [3]. This explains the high incidence of renal artery involvement and hypertension in our patients. In a study of Indian hypertensive patients, TA was responsible for 75 per cent of patients of renovascular hypertension [9]. Unequal pulses, hypertension and arterial bruits were common clinical features. Tenderness over the arteries was not

seen in any case, possibly due to late presentation of our cases. Systemic features were present in only 23.3 per cent cases. Six patients were initially being treated as hypertension and were diagnosed later as TA on detailed evaluation. This is important, as the use of angiotensin convertase inhibitors to control blood pressure, may be catastrophic in such patients. A high index of suspicion for TA in young hypertensives is necessary in our country. A higher incidence of neurological features (26.6%) was seen. On analysis of odds ratio, raised ESR correlated poorly with systemic features. Histologically active disease has been documented in patients without clinical evidence of activity and with normal ESR [6]. The disease runs a protracted course with mortality attributable to hypertension or its effect on heart or brain. Prednisolone in the dose of 1-2 mg/kg body weight results in amelioration of symptoms and improves prognosis in about 50 per cent of the patients with active disease [6]. Maintenance dose of steroids is required for prolonged period. In the remaining glucocorticoid resistant cases, cyclophosphamide and azathioprine with or without low dose steroids are beneficial. More recently, low dose weekly methotrexate with or without steroids has shown promise [10]. When significant occlusive lesions have already occurred, percutaneous transluminal angioplasty or arterial reconstruction is required to limit the effects of hypoperfusion in various organs. Restenosis due to fibrotic healing process or from continued inflammation may be as high as 25 per cent. With the use of self-expanding metallic stents these are likely to come down in future.

TA should be suspected in young patients presenting with hypertension or stroke in our country. Unequal pulses or blood pressure in all four limbs and arterial bruits must be carefully looked for in these patients.

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