Clin Rheumatol (2010) 29:973–981
DOI 10.1007/s10067-010-1496-1

ORIGINAL ARTICLE

Takayasu's arteritis: clinical features and outcomes of 125 patients in China

Xiao-Liang Cong · Sheng-Ming Dai · Xiang Feng · Zhi-Wei Wang · Qing-Sheng Lu · Liang-Xi Yuan · Xian-Xian Zhao · Dong-Bao Zhao · Zai-Ping Jing

Received: 1 March 2009 / Revised: 27 December 2009 / Accepted: 20 May 2010 / Published online: 30 June 2010

Abstract Takayasu's arteritis (TA) is a chronic inflammation that frequently involves the aorta and its major branches. The clinical features of TA vary in different ethnic populations. The objective of this study is to characterize the clinical features, angiographic findings, and response to treatment of patients with TA in Changhai Hospital, Shanghai, China. The hospital records of 125 patients diagnosed with TA were retrospectively evaluated. Eighty patients were followed for a median duration of 36 months. Females (86.4%) were most frequently affected. The mean age at onset was 26.9 years. Constitutional symptoms were present in only 38.4% of patients. The most common clinical finding was pulse deficit. Histological findings from 12 clinically inactive patients showed active lesions in 58.3%. Angiographic classification showed that type I was the most common, followed by type V and IV. Type I was more common in adult patients than in pediatric patients. Although immunosuppressive treatment induced remission in most patients, over 90% of those who achieved later remission relapsed. Both bypass procedures and angioplasty showed high rates of initial success, but restenosis occurred in 34.7% of bypass procedures and 77.3% of angioplasty procedures. Eight patients died during the follow-up period with the main cause of death being congestive heart failure. Constitutional symptoms were not frequent in our study. Correlation between the clinical assessment of disease activity and histologic findings is often poor in TA. Angiographic findings showed that type I was the most common in our study. Over the longer term, the outcomes of revascularization were superior to angioplasty.

Keywords Age · China · Clinical characteristic · Takayasu's arteritis · Treatment

X.-L. Cong · S.-M. Dai (✉) · D.-B. Zhao
Department of Rheumatology and Immunology,
Changhai Hospital, Second Military Medical University,
174 Changhai Road,
Shanghai 200433, People's Republic of China
e-mail: dsm@medmail.com.cn

X. Feng · Q.-S. Lu · L.-X. Yuan · Z.-P. Jing (✉)
Department of Vascular Surgery, Changhai Hospital,
Second Military Medical University,
174 Changhai Road,
Shanghai 200433, People's Republic of China
e-mail: jingzp@xueguan.net

Z.-W. Wang
Department of Orthopedics, Changhai Hospital,
Second Military Medical University,
174 Changhai Road,
Shanghai 200433, People's Republic of China

X.-X. Zhao
Department of Cardiology, Changhai Hospital,
Second Military Medical University,
174 Changhai Road,
Shanghai 200433, People's Republic of China

Introduction

Takayasu's arteritis (TA) is a chronic condition, mainly involving the aorta and its main branches, as well as coronary and pulmonary arteries. The disease has a prominent female preponderance and manifests in the second and third decades of life. TA is now regarded as a worldwide entity, although a higher incidence exists in Southeast Asia, South Africa, and Latin America. The annual incidence of TA was estimated as 2.6 per million in North America [1] and 1.2 per million in Japan [2]. Depending on the location of the affected vessels and the

severity of the vascular inflammation, the clinical presentation of TA may range from asymptomatic disease as a result of diminished pulse to fatal cerebrovascular accident. Reports from different geographical areas suggested heterogeneity in disease expression in various ethnic populations. The aim of the present study was to evaluate the clinical, laboratory, and radiological features and outcomes of TA patients in our hospital. Furthermore, we sought to compare the clinical and angiographic features between juvenile and adult patients.

Patients and methods

Patients

Over a 15-year period from 1993 to 2008, hospital records of 125 patients diagnosed with TA in Changhai Hospital (a hospital of the Second Military Medical University, Shanghai, China) were retrospectively evaluated. Most of the patients were from East China.

Criteria for diagnosis and disease activity

The diagnosis was made according to the classification criteria of the American College of Rheumatology (ACR) for TA [3]. Other causes of large-vessel abnormalities were excluded based on clinical criteria and, where appropriate, on serologic and imaging studies. These included inflammatory aortitis (such as tuberculosis, syphilis, Buerger, Behcet, Cogan, and Kawasaki diseases), developmental anomalies (such as the Marfan syndrome), and other aortic abnormalities (such as neurofibromatosis and radiation fibrosis). Disease activity was judged according to the National Institutes of Health (NIH) criteria [4]. Angiographic classification was made according to the criteria suggested by Numano's group [5]. Histological findings of biopsy specimens obtained from patients during bypass procedures were divided into two types: (1) active lesions, characterized by perivascular mononuclear cell infiltrates in the vasa vasorum, and (2) fibrotic lesions, characterized by perivascular fibrosis and fibrous thickening of media.

Statistical methods

Statistical data were analyzed using the SPSS version 13.0 software. The comparisons of clinical and angiographic findings between groups were performed using the Chi-square and Fisher's exact test. Diagnostic delay time for studied groups was compared using the Wilcoxon's rank sum test. P values less than 0.05 were considered statistically significant.

Results

Of the 125 patients, 108 were female with a male/female ratio of 1:6.35. Mean age at onset was 26.9 years (6–65 years), and 90.4% of patients presented before age 40 years. Thirty-one patients were juveniles (≤18 years) at onset, with a mean age of 13.7 years (6–18 years). The male/female ratio was 1:3.4 in juveniles and 1:8.4 in adults. Median diagnostic delay, defined as median time elapsed from the onset of symptoms to diagnosis, was 19 months (0.5–160 months) for the entire group. The median delay time was 20 months in pediatric patients and 15.5 months in adult patients (P=0.984).

Clinical features

Constitutional symptoms

Constitutional symptoms were present in over one-third of the patients (Table 1). Of these symptoms, malaise and fever were most common. Constitutional symptoms were more common in adult patients compared to pediatric patients (P=0.037, 95% confidence interval (CI), 1.041–6.759; Table 1).

Vascular findings

The most frequent clinical manifestation was pulse deficit on the extremities (Table 1). Pulse deficit was more common in adult patients than in pediatric patients (P=0.005, 95% CI 1.385–7.666; Table 1). Hypertension was present in almost two-thirds of patients, attributing to renal artery stenosis in 53 patients and to coarctation of the aorta in eight patients. Vascular bruits were present most often over the carotid arteries (46.4%) and were less common in the abdomen (26.4%) and femoral (4.8%) regions. Intermittent claudication was more common in the upper limbs (35.2%) compared to the lower limbs (15.2%). None of the patients with mesenteric artery or celiac trunk stenosis developed gastrointestinal symptoms.

Cardiac findings

The most common cardiac finding was aortic valvular regurgitation (20%) which was secondary to aortic root dilation. Aortic valve replacement was required in one patient with severe aortic root dilatation. Cardiac enlargement and ischemic heart disease were found in 31 and 12 patients, respectively. Nine patients presented with symptoms of angina pectoris, and myocardial infarction occurred in one patient. Congestive heart failure, often due to hypertension and aortic valvular regurgitation, occurred in 13.6% of patients.

Table 1 Main characteristics of patients and comparisons between juveniles and adults at time of diagnosis

	Total *N* (%)	Juveniles *N* (%)	Adults *N* (%)	*P*
N	125	31	94	
Sex				
F	108	24 (77.4)	84 (89.4)	0.128
M	17	7 (22.6)	10 (10.6)	0.128
Constitutional findings	48 (38.4)	7 (22.6)	41 (43.6)	0.037
Fever	22 (17.6)	5 (16.1)	17 (18.1)	0.804
Malaise	32 (25.6)	5 (16.1)	27 (28.7)	0.164
Joint pain	11 (8.8)	2 (6.5)	9 (9.6)	0.730
Night sweat	5 (4.0)	1 (3.2)	4 (4.3)	1.000
Weight loss	3 (2.4)	0	3 (3.2)	0.574
Vascular findings				
Pulse deficit	89 (71.2)	16 (51.6)	73 (77.7)	0.005
Claudication	55 (44.0)	13 (41.9)	42 (44.7)	0.789
Vascular bruit	78 (62.4)	16 (51.6)	62 (66.0)	0.153
Hypertension	82 (65.6)	23 (74.2)	59 (62.8)	0.245
Asymmetric blood pressure	42 (33.6)	8 (25.8)	34 (36.2)	0.289
Cardiac findings				
Dyspnea on exertion	25 (20.0)	7 (22.6)	18 (19.1)	0.679
Palpitation	11 (8.8)	1 (3.2)	10 (10.6)	0.290
Angina	9 (7.2)	0	9 (9.6)	0.111
Left ventricular hypertrophy	29 (23.2)	6 (19.4)	23 (24.5)	0.559
Neurologic findings				
Headache	32 (25.6)	11 (35.5)	21 (22.3)	0.146
Dizziness	63 (50.4)	13 (41.9)	50 (53.2)	0.277
Visual disturbance	42 (33.6)	8 (25.8)	34 (36.2)	0.289
Ablepsia	12 (9.6)	3 (9.7)	9 (9.6)	1.000
Syncope	20 (16.0)	3 (9.7)	17 (18.1)	0.398
Retinopathy	20 (16.0)	5 (16.1)	15 (16.0)	1.000
Laboratory findings				
Elevated ESR	62 (49.6)	5 (16.1)	57 (60.6)	0.000
Elevated CRP	61/101	20/31	41/94	
Anemia	39 (31.2)	6 (19.4)	33 (35.1)	0.101
Angiographic classification				
Type I	50	6 (19.4)	44 (46.8)	0.007
Type IIa	6	1 (3.2)	5 (5.3)	1.000
Type IIb	2	2 (6.5)	0	0.060
Type III	3	2 (6.5)	1 (1.1)	0.152
Type IV	26	10 (32.3)	16 (17.0)	0.070
Type V	38	10 (32.3)	28 (29.8)	0.795

Neurologic findings

Dizziness was the most common neurological symptom (Table 1). Visual disturbances, such as scotoma, blurring, or diplopia, were most often bilateral. Typical retinopathy secondary to hypertension or ischemia was found in 16% of the patients. Permanent ablepsia attributed to TA was most often monocular. Cerebrovascular accidents occurred in 20 patients (18 with cerebral infarction, 2 with cerebral hemorrhage).

Angiographic findings

All patients underwent pan-angiography at the time of diagnosis. A total of 621 lesions were detected in 125 patients (Table 2). A predominance of stenosis or occlusion

Table 2 Angiographic findings in patients with Takayasu's arteritis (N=125)

Artery	Total abnormalities	Stenosis	Dilatation	Aneurysm
Aorta				
Aortic root	10	4	6	0
Aortic arch	11	8	3	0
Thoracic aorta	17	15	2	1
Abdominal aorta	55	52	6	2
Innominate	28	28	0	0
Common carotid				
Left only	33	33	0	0
Right only	6	6	1	0
Bilateral	41	41	0	0
Subclavian				
Left only	42	42	1	0
Right only	4	4	0	0
Bilateral	40	40	1	0
Vertebral				
Left only	16	16	0	0
Right only	6	6	0	0
Bilateral	18	17	2	0
Celiac trunk	17	17	1	1
Superior mesenteric	19	18	1	0
Inferior mesenteric	2	2	0	0
Renal				
Left only	14	14	1	0
Right only	13	13	1	0
Bilateral	26	26	0	0
Common iliac				
Left only	4	4	0	0
Right only	1	0	0	1
Bilateral	9	9	1	1
Common femoral				
Left only	2	2	0	0
Right only	4	4	0	0
Bilateral	7	7	0	0
Pulmonary	10	7	3	0
Coronary	5	5	0	0

lesions was present in 93.4% of involved arteries; other lesions were less common (5.5% with dilatation, 1.1% with aneurysm). Based on the angiographic findings, type I (40%), type IV (20.8%), and type V (30.4%) were common, whereas type IIa (4.8%), type IIb (1.6%), and type III (2.4%) were less common (Table 3). Compared with pediatric patients, type I was more common in adult patients (P=0.007, 95% CI 1.378–9.758; Table 1). Among the nine cases with chest pain, five showed involvement of coronary arteries. All patients had ostial left main coronary artery involvement. Pulmonary hypertension was diagnosed in 14 patients by echocardiography, and ten of these patients had pulmonary artery involvements confirmed by angiography (stenosis in four, occlusion in three, and dilatation in three patients).

Laboratory and histologic findings

The median erythrocyte sedimentation rate (ESR) was 10 mm/h (range 2–123) in pediatric patients and 25 mm/h (range 1–140) in adult patients. Elevated ESR was more common in adult patients than in pediatric patients (P=0.000, 95% CI 2.824–22.726). Glomerular filtration rate measured by emission computed tomography was performed in patients with

Table 3 Angiographic characteristics of patients with Takayasu's arteritis in China compared with other geographic areas

Type	China *N* (%)	Thailand [23] *N* (%)	Korea [21] *N* (%)	Brazil [24] *N* (%)	Colombia [19] *N* (%)	India [25] *N* (%)	Japan [25] *N* (%)	Iran [14] *N* (%)
I	50 (40)	0	39 (36.1)	4 (13.3)	12 (34.3)	7 (6.9)	19 (24.1)	8 (10.3)
IIa	6 (4.8)	0	3 (2.8)	1 (3.3)	4 (11.4)	1 (1.0)	9 (11.4)	12 (15.4)
IIb	2 (1.6)	7 (11.1)	5 (4.6)	0	2 (5.7)	6 (5.9)	8 (10.1)	0
III	3 (2.4)	2 (3.2)	8 (7.4)	1 (3.3)	0	3 (2.9)	0	14 (17.9)
IV	26 (20.8)	12 (19.0)	17 (15.7)	4 (13.3)	7 (20.0)	29 (28.4)	1 (1.3)	14 (17.9)
V	38 (30.4)	42 (66.7)	36 (33.3)	20 (66.7)	10 (28.6)	56 (54.9)	42 (53.2)	30 (38.5)
Total	125	63	108	30	35	102	79	78

renal arteries affected. In 48 patients with renal artery involvement, over two-thirds (67.7%) of them had impaired function.

Arterial biopsy specimens were obtained from 12 patients with assumed clinically inactive disease. However, active lesions were observed in seven patients, and fibrotic lesions were observed in five patients. None of the seven patients with histologically active disease showed an elevated ESR.

Treatment and outcomes

Eighty patients were followed for a median duration of 36 months (3–180 months). In this group, 85% were female, and the median age at onset was 26 years (range 6–62 years). The demographic characteristics of the follow-up group were similar to that of the entire group of 125 patients.

Medical therapy

During the disease course, glucocorticoids (GC) were prescribed to 58 patients (72.5%). Sixteen patients required cytotoxic agents in addition to GC. Of them, a cytotoxic agent was added in ten patients because of failure to induce remission with GC alone and in six patients due to the inability to taper GC dosages once the disease was controlled. These cytotoxic agents included methotrexate (N=4), azathioprine (N=2), mycophenolate mofetil (N=2), and cyclophosphamide (N=8). Remission of any duration was achieved in 57 of 58 patients. One patient continued to have active disease despite therapy with GC and other immunosuppressive agents.

Fifty-two of the 57 patients (91.2%) who had achieved remission experienced at least one relapse. Only five of the 16 patients treated with a combination of cytotoxic agents and GC experienced no relapse.

Percutaneous balloon angioplasty

During the follow-up period, 20 patients received angioplasty procedures. Vascular procedures were only performed when disease was judged to be quiescent. Thirteen of the 20 patients had active disease and received strict perioperative GC during vascular procedures. Other seven patients in quiescent phase were only treated with preventive GC. During the disease course, 25 angioplasty procedures (in 20 patients) were performed, and 22 (ten with stents) were initially successful. These procedures included carotid angioplasty (three procedures, two with stents), renal angioplasty (15 procedures, 6 with stents), aortic angioplasty (two procedures, none with stents), subclavian angioplasty (one procedure with stent), and coronary angioplasty (one procedure with stent). Three procedures were unsuccessful. In one patient, failure was due to the inability to place a stent in the vessel. In the other two patients, the guide wire could not cross the occluded subclavian artery lesion, and angioplasty was terminated. Restenosis occurred in 17 (77.3%) of the 22 initially successful angioplasty procedures with or without stents. Fourteen of the 17 restenotic sites (82.4%) developed in less than 1 year.

Vascular reconstruction

The indications for vascular interventions included renovascular hypertension, extremity claudication, cerebrovascular ischemia, and cardiac ischemia. Vascular bypass was performed in 36 patients (49 procedures in total). Twenty-three of the 36 patients had active disease and received perioperative GC during procedures. Other 13 patients in quiescent phase were treated with preventive GC. Mean duration of symptoms before operation was 21.4 months (1–120 months). Two patients died after the procedures. The cause of death was acute pericardial hemorrhage and cerebral hemorrhage, respectively. Postoperative complications occurred in four patients (one patients with infection, two patients with strokes, one patient had in-hospital carotid graft occlusion). Synthetic grafts were used for 24 bypass procedures. Autologous vessel grafts (saphenous vein) were often selected for a bypass of stenotic lesions of the renal and coronary arteries and were used for 25 bypass

procedures. Restenosis or thrombosis was identified in 34.7% of these procedures. Ten of the 24 synthetic grafts (41.6%) developed restenosis or thrombosis during the follow-up. Seven of the 25 (28%) saphenous veins were followed by anastomotic stenosis.

Morbidity and mortality

Follow-up of 80 patients revealed that congestive heart failure developed in four patients, aortic regurgitation in three patients, renal failure in two patients, fundus hemorrhage in one patient, and cerebral infarction in one patient. Thirty-two of the 80 (40%) patients had impaired function of routine daily life owing to vascular claudication, congestive heart failure, or residual hemiplegia. Eight patients died during the follow-up period (Table 4).

Discussion

The average age of 26.9 years at onset is consistent with the previous reports from the USA and other Asian countries [6–10]. This finding is in contrast to the earlier age reported in Latin America [11] and the later age in Europe [12]. A significant proportion (9.6%) of our patients who otherwise fulfilled the diagnostic criteria was older than 40 years at disease onset. This finding contradicts the current ACR classification criteria for TA including an "age at disease onset of less than 40 years" [3].

The female-to-male ratio among patients with TA varies from 1.2:1 to 9.4:1 [11, 13–16]. We observed a female/male ratio of 6.35:1, which is consistent with an Italian study with the ratio of 7:1 [15]. This finding contradicts the belief that the female-to-male ratio increases when moving toward the East [17]. In contrast to adult patients, the female preponderance in pediatric patients (male/female ratio=1:3.4) seemed less obvious. This may be attributed to the hormonal level discrepancy between adult and pediatric patients. This finding is further supported by necrosis and inflammatory reactions simulating TA and atrophy of muscular layers in aorta of rabbits treated with estrogens [18].

There is often a delay in diagnosis of TA from the onset of first symptoms. This delay can be months or even years. The NIH study suggested that the delay in diagnosis was longer in juveniles (<20 years), being up to four times that of adult patients [4]. Vanoli et al. [15] also found that age at onset of <15 years was associated with a higher probability of a delay in diagnosis. However, we achieved different results. Data looking at patients aged less than 18 years did not show a significantly longer delay in diagnosis. This discrepancy presumably relates to the difference in disease incidence, resulting in a difference in awareness.

TA is relatively rare and may have an insidious and nonspecific presentation. A prompt diagnosis of TA is difficult, because of the failure to recognize the disease. Constitutional signs and symptoms indicative of an inflammatory disease occurred in only 38.4% of our patients. Similar results have been reported in the USA [4] and Colombia [19]. This finding failed to support the notion that the early phase of TA is frequently characterized by general symptoms of systemic inflammation. Although vascular ischemic symptoms, e.g., bruit or diminished pulses, are common in patients with TA and should guide the physician to suspect the disease, ischemic symptoms present at a stage when stenotic or occlusive lesions have already occurred. In our study, the incidence of leading diagnostic clues such as constitutional symptoms or certain ischemic symptoms was lower in juvenile compared to adult patients. Our findings were similar to the findings from the USA [4]. This may explain the long delay in diagnosis of TA in American and Italian juvenile patients [4, 15].

Systemic hypertension was more prevalent in our study compared to studies in Colombia [19], USA [4], and Korea

Table 4 Clinical and angiographic features and cause of death (N=8)

Number	Sex	Age at onset	Delay time (years)[a]	Disease Activity	Angiographic type	Serious complication	Cause of death
1	F	13	10	Active	IIb	RVH, CHF, VHD	CHF
2	F	15	7	Active	V	RVH, CHF, VHD	CHF
3	F	43	11	Active	IV	RVH, CHF, VHD, CRF	CHF
4	F	26	10	Active	I	RVH, VHD	CHF
5	F	22	9	Stable	IV	RVH	CVA
6	M	6	4	Active	IV	CHF, CVA, RVH	CHF
7	M	17	7	Stable	IV	CHF, VHD, CRF, RVH	CRF
8	F	21	35	Active	V	IHD, CVA	AMI

[a] Delay time from diagnosis to death

RVH renovascular hypertension, *CHF* chronic heart failure, *VHD* valvular heart disease, *CRF* chronic renal failure, *CVA* cerebrovascular accident, *IHD* ischemic heart disease, *AMI* acute myocardial infarction

[10]. Hypertension is frequently unrecognized and untreated. Since both subclavian and axillary arteries are often involved, hypertension cannot be adequately monitored using conventional blood pressure cuffs. Clinicians should take the blood pressure in extremity without arteries affected as results on the basis of angiography. Central blood pressure determination is not feasible in daily clinical practice and should always be obtained in patients with extremity peripheral vessel stenosis. Hypertension was most often associated with renal artery stenosis. In hypertensive patients without the evidence of renal artery stenosis, the marked narrowing of aorta, abnormal vascular compliance, dysfunction baroreceptors, and long-term prednisone may have also contributed to hypertension [4, 14]. Surgical correction of renal artery stenosis has, for the most part, focused on the use of bypass grafts and is usually an effective means of decreasing or eliminating hypertension.

Assessment of disease activity is crucial to improve treatment outcomes in TA. However, the current laboratory parameters from patients with TA lack specificity and sensitivity. In our study, biopsy data demonstrated active inflammation in 58.3% of patients thought to be in clinical remission. None of these patients had an elevated ESR. ESR was therefore an unreliable surrogate marker for assessing the disease activity in TA. In contrast, some studies have found ESR to be a reliable and useful marker to determine the status of disease activity [20, 21]. Acute-phase reactants are not sufficient to rely on as a definite measure of quiescent disease. However, elevated acute-phase reactants should encourage further evaluation to identify additional evidence of disease activity.

Most of the patients with TA had extensive disease of the aorta and its branches, and the predominant anatomic lesion was stenosis or occlusion (93.4%). This finding was similar to the angiographic features reported in other countries [6, 8, 21, 22]. However, the involvement of arteries revealed variations in different countries. Table 3 compares arterial involvement of cases from China with those from other countries. In our study, type I was the most frequent, suggesting that the patients have relatively favorable vascular lesions. This is because type I represents a focal vascular lesion confined to the branches of the aortic arch. A similar pattern of vascular involvement was also observed in Korea and Colombia. In contrast, type V which is a generalized type with combined features of the other types was the most frequent in Thailand, Brazil, India, and Japan and has been less commonly observed in our study. Type IV which involves only the abdominal aorta and/or the renal arteries was less common in Japan, whereas type IV prevails in Thai, Indian, Colombian, Iranian and our patients. The variations in patterns of vascular involvement in different areas indicate that ethnic factors may play an important role in pathogenesis of TA. Compared with pediatric patients, the involvement of aortic arch branches (type I) was more common in adults and may explain the lower incidence of vascular ischemic symptoms such as diminished pulse, claudication, or bruit in pediatric patients. The reason for the angiographic difference between pediatric and adult patients is unknown. It may have implications for future study in the field of age-related genes in the etiopathogenesis and distribution of vascular lesions in TA.

Coronary involvement is detected in approximately 10% to 30% of TA cases [26]. In our study, we performed coronary angiography in nine patients with chest pain. Of these patients, five had coronary arteries involvement. All of the patients had ostial left main coronary artery involvement, which was also a prevalent feature in other studies [27, 28]. This finding was supposed to be secondary to the inflammation of intima and the contraction of the fibrotic media and adventitia from the ascending aorta. Pulmonary arteritis is a feature unique to TA and is not found in other forms of aortitis. An abnormal pulmonary angiography is present in as high as 30–74% of patients [29, 30]. It is likely that failure to routinely perform pulmonary angiography has resulted in an underestimation of its frequency in present patients.

GC constitutes the first line of treatment for active TA and is an effective palliative agent for most patients. In this study, a total of 48 of 58 patients treated with GC alone achieved disease remission. In other series, the successful response to GC ranged from 20% to 100% [4, 11, 31, 32]. Although remission was achieved in most patients, GC-resistant patients and relapses during GC tapering were also common in our study. Therefore, cytotoxic agents were added to GC in 16 patients with the aim to further control disease activity. The addition of cytotoxic agents often allowed for further reduction in the GC dose and was partly effective in corticoid-resistant cases. However, relapses remained common despite the use of adjunctive immunosuppressive therapies. Additionally, medical treatments did not reverse established vascular lesions in this study. Therefore, for patients with TA, disease control may be a more realistic therapeutic goal than eradication.

Stenotic or occlusive lesions are usually not reversible by medical treatment. If these lesions are hemodynamically significant, angioplasty or revascularization may be required. Initial studies that examined the outcomes of percutaneous balloon angioplasty (PTA) in TA seemed promising. Tyagi et al. [33] reported an initial success rate of 89% for PTA in the treatment of 45 TA patients. The patency rate was 79% at a mean follow-up of 43 months. Similar findings were also reported by Sharma and colleagues [34]. However, our results revealed a less optimistic view of PTA in TA. In our study, 25 angioplasty

procedures were performed with an initial success rate of 88% (22/25). Restenosis occurred in 77.3% (17/22) of the procedures that were initially successful. These findings are supported by longitudinal data from two American series [4, 6]. Such results could be explained by the nature of vessel lesions in TA, which are usually long, fibrotic, and nearly or completely occluded. Thus, they may be less amenable to successful dilation. In addition, the trauma of dilatation could also lead to enhanced myointimal proliferation. The relationship between the outcomes of PTA and angiotype was not significant. Outcomes of arterial bypass procedures in TA have demonstrated better sustained patency compared to PTA procedures. In our study, 49 bypass procedures were performed in 36 patients. Overall, 34.7% of these procedures were followed by restenosis or thrombosis, which may or may not have been hemodynamically significant. On average, 20% to 35% of restenosis or occlusion was reported in other series on long-term follow-up [6, 35, 36]. Fields et al. [37] reported that patients with active disease requiring operations were more likely to develop thrombosis or restenosis. It is therefore recommended that surgical interventions be performed when disease is judged to be quiescent. Our finding that 58.3% of arterial biopsy specimens from the patients with assumed clinically quiescent disease showed active vasculitis underscores the poor correlation between clinical assessment and disease activity. This histologic evidence of covert active inflammation confirms the argument for perioperative immunosuppressive therapy for patients with TA.

In summary, constitutional symptoms were not common in our study. Generally, clinical presentations were similar in both pediatric and adult patients. Involvement of aortic arch branches was more common in adult patients compared to pediatric patients. The ESR was not a reliable surrogate marker of disease activity. Repeated and, at times, prolonged courses of immunosuppressive therapy are often required due to the relapsing nature of TA. Although ischemic symptoms significantly palliated after angioplasty or bypass of stenotic vessels, restenosis was common.

Disclosures None

References

1. Hall S, Barr W, Lie JT, Stanson AW, Kazmier FJ, Hunder GG (1985) Takayasu arteritis. A study of 32 North American patients. Medicine (Baltimore) 64:89–99
2. Nasu T (1975) Takayasu's truncoarteritis in Japan. A statistical observation of 76 autopsy cases. Pathol Microbiol (Basel) 43:140–146
3. Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy SM, Fauci AS, Leavitt RY, Lie JT, Lightfoot RW Jr et al (1990) The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum 33:1129–1134
4. Kerr GS, Hallahan CW, Giordano J, Leavitt RY, Fauci AS, Rottem M, Hoffman GS (1994) Takayasu arteritis. Ann Intern Med 120:919–929
5. Hata A, Noda M, Moriwaki R, Numano F (1996) Angiographic findings of Takayasu arteritis: new classification. Int J Cardiol 54 (Suppl):155–163
6. Maksimowicz-McKinnon K, Clark TM, Hoffman GS (2007) Limitations of therapy and a guarded prognosis in an American cohort of Takayasu arteritis patients. Arthritis Rheum 56:1000–1009
7. Ishikawa K (1986) Patterns of symptoms and prognosis in occlusive thromboaortopathy (Takayasu's disease). J Am Coll Cardiol 8:1041–1046
8. Subramanyan R, Joy J, Balakrishnan KG (1989) Natural history of aortoarteritis (Takayasu's disease). Circulation 80:429–437
9. Ishikawa K, Maetani S (1994) Long-term outcome for 120 Japanese patients with Takayasu's disease. Clinical and statistical analyses of related prognostic factors. Circulation 90:1855–1860
10. Park YB, Hong SK, Choi KJ, Sohn DW, Oh BH, Lee MM, Choi YS, Seo JD, Lee YW, Park JH (1992) Takayasu arteritis in Korea: clinical and angiographic features. Heart Vessels Suppl 7:55–59
11. Lupi-Herrera E, Sánchez-Torres G, Marcushamer J, Mispireta J, Horwitz S, Vela JE (1977) Takayasu's arteritis. Clinical study of 107 cases. Am Heart J 93:94–103
12. Waern AU, Andersson P, Hemmingsson A (1983) Takayasu's arteritis: a hospital-region based study on occurrence, treatment and prognosis. Angiology 34:311–320
13. Deutsch V, Wexler L, Deutsch H (1974) Takayasu's arteritis. An angiographic study with remarks on ethnic distribution in Israel. Am J Roentgenol Radium Ther Nucl Med 122:13–28
14. Sheikhzadeh A, Tettenborn I, Noohi F, Eftekharzadeh M, Schnabel A (2002) Occlusive thromboaortopathy (Takayasu disease): clinical and angiographic features and a brief review of literature. Angiology 53:29–40
15. Vanoli M, Daina E, Salvarani C, Sabbadini MG, Rossi C, Bacchiani G, Schieppati A, Baldissera E, Bertolini G (2005) Takayasu's arteritis: a study of 104 Italian patients. Arthritis Rheum 53:100–107
16. Koide K (1992) Takayasu arteritis in Japan. Heart Vessels Suppl 7:48–54
17. Numano F, Okawara M, Inomata H, Kobayashi Y (2000) Takayasu's arteritis. Lancet 356:1023–1025
18. Numano F, Shimamoto T (1971) Hypersecretion of estrogen in Takayasu's disease. Am Heart J 81:591–596
19. Cañas CA, Jimenez CA, Ramirez LA, Uribe O, Tobón I, Torrenegra A, Cortina A, Muñoz M, Gutierrez O, Restrepo JF, Peña M, Iglesias A (1998) Takayasu arteritis in Colombia. Int J Cardiol 66(Suppl 1):73–79
20. Mwipatayi BP, Jeffery PC, Beningfield SJ, Matley PJ, Naidoo NG, Kalla AA, Kahn D (2005) Takayasu arteritis: clinical features and management: report of 272 cases. ANZ J Surg 75:110–117
21. Park MC, Lee SW, Park YB, Chung NS, Lee SK (2005) Clinical characteristics and outcomes of Takayasu's arteritis: analysis of 108 patients using standardized criteria for diagnosis, activity assessment, and angiographic classification. Scand J Rheumatol 34:284–292
22. Jain S, Kumari S, Ganguly NK, Sharma BK (1996) Current status of Takayasu arteritis in India. Int J Cardiol 54(Suppl):111–116
23. Suwanwela N, Piyachon C (1996) Takayasu arteritis in Thailand: clinical and imaging features. Int J Cardiol 54(Suppl):117–134

24. Sato EI, Lima DN, Espirito Santo B, Hata F (2000) Takayasu arteritis. Treatment and prognosis in a university center in Brazil. Int J Cardiol 75(Suppl 1):163–166
25. Moriwaki R, Noda M, Yajima M, Sharma BK, Numano F (1997) Clinical manifestations of Takayasu arteritis in India and Japan-new classification of angiographic findings. Angiology 48:369–379
26. Rav-Acha M, Plot L, Peled N, Amital H (2007) Coronary involvement in Takayasu's arteritis. Autoimmun Rev 6:566–571
27. Brueren BR, Ernst JM, Suttorp MJ, ten Berg JM, Rensing BJ, Mast EG, Bal ET, Six AJ, Plokker HW (2003) Long term follow up after elective percutaneous coronary intervention for unprotected non-bifurcational left main stenosis: is it time to change the guidelines? Heart 89:1336–1339
28. Punamiya K, Bates ER, Shea MJ, Muller DW (1997) Endoluminal stenting for unprotected left main stenosis in Takayasu's arteritis. Catheter Cardiovasc Diagn 40:272–275
29. Liu YQ, Jin BL, Ling J (1994) Pulmonary artery involvement in aortoarteritis: an angiographic study. Cardiovasc Interv Radiol 17:2–6
30. Sharma S, Kamalakar T, Rajani M, Talwar KK, Shrivastava S (1990) The incidence and patterns of pulmonary artery involvement in Takayasu's arteritis. Clin Radiol 42:177–181
31. Ishikawa K (1991) Effects of prednisolone therapy on arterial angiographic features in Takayasu's disease. Am J Cardiol 68:410–413
32. Fraga A, Mintz G, Valle L, Flores-Izquierdo G (1972) Takayasu's arteritis: frequency of systemic manifestations (study of 22 patients) and favorable response to maintenance steroid therapy with adrenocorticosteroids (12 patients). Arthritis Rheum 15:617–624
33. Tyagi S, Gambhir DS, Kaul UA, Verma P, Arora R (1996) A decade of subclavian angioplasty: aortoarteritis versus atherosclerosis. Indian Heart J 48:667–671
34. Sharma S, Saxena A, Talwar KK, Kaul U, Mehta SN, Rajani M (1992) Renal artery stenosis caused by nonspecific arteritis (Takayasu disease): results of treatment with percutaneous transluminal angioplasty. AJR Am J Roentgenol 158:417–422
35. Liang P, Tan-Ong M, Hoffman GS (2004) Takayasu's arteritis: vascular interventions and outcomes. J Rheumatol 31:102–106
36. Weaver FA, Kumar SR, Yellin AE, Anderson S, Hood DB, Rowe VL, Kitridou RC, Kohl RD, Alexander J (2004) Renal revascularization in Takayasu arteritis-induced renal artery stenosis. J Vasc Surg 39:749–757
37. Fields CE, Bower TC, Cooper LT, Hoskin T, Noel AA, Panneton JM, Sullivan TM, Gloviczki P, Cherry KJ Jr (2006) Takayasu's arteritis: operative results and influence of disease activity. J Vasc Surg 43:64–71