

TAKAYASU'S ARTERITIS: THE RETROSPECTIVE ANALYSIS OF PATIENTS FROM THE URAL POPULATION

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Takayasu's arteritis (TA) is a rare disease that can be overlooked during the first visit to a GP, rheumatologist, or any other medical specialist due to a variety of its symptoms. The aim of this study was to describe the clinical presentation and the course of patients with TA residing in the Middle Ural. A retrospective analysis was conducted using the medical records of 183 patients treated at the Sverdlovsk Regional Clinical Hospital 1 from 1979 through 2018. The male to female ratio was 1:3. The mean age was 33.5 years for women and 35.2 for men. The most frequently involved arteries were subclavian (101 cases; 55%), carotid (98 cases; 53%) and renal (77 cases; 42%). Type V was the most common angiographic type. Arterial stenosis was present in 94 (51%) patients. Sixty-six patients received surgical interventions. Of all patients included in the analysis, 31 died. The observed 5-year survival was 92%, 10-year survival, 90% and 15-year survival, 80%. Seventy-two patients (39%) developed major adverse cardiovascular events (MACE), including myocardial infarction, ischemic stroke, and thrombosis of large arteries/veins. The clinical presentation of TA may vary in different geographical regions.

Keywords: Takayasu's arteritis, clinical symptoms, renal artery**Author contribution:** all authors participated in conceiving and planning the study, processing the data, discussing the results, and writing the manuscript. Borodina IE and Salavatova GG collected the medical records for the electronic database.**Compliance with ethical standards:** the study was approved by the Ethics Committee of Ural State Medical University (Protocol No. 9 dated November 23, 2018).✉ **Correspondence should be addressed:** Irina E. Borodina
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АРТЕРИИТ ТАКАЯСУ: РЕЗУЛЬТАТЫ РЕТРОСПЕКТИВНОГО АНАЛИЗА ПАЦИЕНТОВ УРАЛЬСКОЙ ПОПУЛЯЦИИ

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Артериит Такаясус (АТ) — редкое заболевание, которое ревматологи, врачи общей практики и другие специалисты могут не распознать при первичном посещении пациента из-за различных клинических проявлений. Целью исследования было оценить клиническую картину и течение АТ у 183 пациентов Среднего Урала. В ретроспективную часть исследования вошли 183 страдающих АТ пациентов, наблюдавшиеся в Свердловской областной клинической больнице № 1 в период с 1979 по 2018 г. Соотношение мужчин и женщин составляло 1 : 3. Средний возраст женщин — 33,5 года, мужчин — 35,2 лет. Наиболее часто были зарегистрированы поражения следующих артерий: подключичной — 101 (55%) случай, сонной — 98 (53%) случаев и почечных — 77 (42%) случаев. Наиболее часто встречался ангиографический тип V, а типичным ангиографическим признаком был артериальный стеноз — 94 (51%). Хирургические вмешательства выполнены 66 пациентам. За период исследования зарегистрирован 31 летальный исход, пятилетняя выживаемость составила 92%; 10-летняя — 90%, а 15-летняя — 80%. У 72 (39%) пациентов развились клинически значимые сердечно-сосудистые события: инфаркт миокарда, ишемический инсульт, тромбоз крупной артерии и венозный тромбоз. В разных географических зонах АТ может иметь широкий спектр клинических проявлений.

Ключевые слова: артериит Такаясус, клинические проявления, почечная артерия**Информация о вкладе авторов:** все авторы принимали участие в планировании работы, статистической обработке, обсуждении результатов, написании и редактировании текста. И. Э. Бородина и Г. Г. Салаватова проводили сбор первичного материала и формирование электронной базы данных.**Соблюдение этических стандартов:** исследование одобрено этическим комитетом ФБГОУ ВО УГМУ Министерства здравоохранения России, протокол № 9 от 23 ноября 2018 г.✉ **Для корреспонденции:** Ирина Эдуардовна Бородина
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Takayasu's arteritis (TA) is a granulomatous vasculitis of the aorta and its major branches. Although TA predominantly affects Asian and South American ethnic groups, rare cases of the disease are reported in other races, too [1]. TA incidence varies from 0.8 to 2.6 cases per 1, 000, 000 adult population depending on the geographical area of residence and ethnicity [2]. At present, no epidemiological data are available on the prevalence of this disease in the Russian Federation. Because of untimely diagnosis and delayed treatment, patients with TA can develop major adverse cardiovascular events (MACE) leading to premature yet preventable death.

At its onset, TA can mimic a wide range of conditions. Specific symptoms that may not be present in the early

stages include asymmetric pulse or blood pressure in the upper extremities accompanied by pronounced hypertension, impaired vision, and abdominal pain.

The aim of this retrospective study was to describe the clinical manifestations, laboratory and radiographic findings, the course and outcome of TA in 183 patients residing in the Middle Ural.

METHODS

Our retrospective cohort study included 183 patients (139 females and 44 males) with verified TA who had received treatment at Sverdlovsk Clinical Hospital No. 1 between 1979

and 2018. The age at diagnosis was 9 to 62 years in females (mean: 33.5; median: 35; 25%–75% IQR: 24–43) and 12 to 59 years in male patients (mean: 35.18; median: 34; 25%–75% IQR: 26.5–42). The duration of the disease was 0.6 to 64 years in women (mean: 12.3 years; median: 10 years; 25%–75% IQR: 4–18) and 0.6 to 32 years in men (mean: 9 years; median: 7 years; 25%–75% IQR: 4–14.5). The time elapsed between the onset of the first clinical symptoms and the established diagnosis was 0.6–54 years in females (mean: 5.7 years; median: 3 years; 25%–75% IQR: 1–7) and 0.6–33 years in men (mean: 6 years; median: 4 years; 25%–75% IQR: 1.5–8) (Table 1).

The primary diagnosis of TA specified in the patients' medical records was verified according to the criteria proposed by the American College of Rheumatology in 1990 [3].

Following the unified protocol, we collected and analyzed demographic data, clinical, laboratory and angiography findings, and information about TA-related surgical interventions. Moriwaki's classification criteria were applied to

describe arterial damage [4]. Because our retrospective study covered a long time period that had seen an evolution in clinical practices and technologies, the imaging modalities used to identify arterial stenosis, occlusion, dilation, dissection, etc. were different and included conventional catheter angiography and/or CTA and/or MRA and/or ultrasonography. Importantly, for every patient, ultrasonography findings were confirmed by at least one contrast-enhanced imaging technique.

To obtain information about the clinical manifestations and angiographic demonstration of the disease in other ethnicities, we searched the PubMed database using *Takayasu's arteritis* as keywords. Similar to the present study, the inclusion criteria used in the PubMed articles were based on ACR 1990 [3]. Among them were: the presence of 3 or more TA symptoms (90% sensitivity, 97.8% specificity), including 40 years at onset, claudication (muscle weakness and pain in the extremities during movement), pulse deficit in one or both brachial arteries, blood pressure difference > 10 mmHg in the brachial arteries,

Table 1. Characteristics of the patients with Takayasu's arteritis included in the study

Characteristics	Patients		
	Females 139 (76%)	Males 44 (24%)	Total n = 183 (100%)
Age at diagnosis, years, Me [25%–75%]	35 [24–43]	34 [26.5–42]	35 [24–43]
Duration of the disease, years, Me [25%–75%]	10 [4–18]	7 [4–14.5]	13.5 [6–20]
Time between the onset of the first symptoms and diagnosis, Me [25%–75%]	3 [1–7]	4 [1.5–8]	3 [1–8]

Table 2. Clinical and laboratory findings

	Patients (n = 183)
Hypertension	98 (53%)
Pain, weakness, numbness, and asymmetric pulse in the upper extremities	89 (49%)
Malaise	87 (47.5%)
Headache	86 (47%)
Fever	65 (35.5%)
Weight loss	44 (24%)
Claudication	40 (22%)
Dizziness	40 (2%)
Blood pressure difference > 10 mmHg	37 (20%)
Chronic abdominal ischemia	36 (20%)
Chest pain	31 (17%)
Pain, weakness, numbness in the lower extremities	26 (14%)
Shortness of breath	16 (8%)
Arthralgias	13 (7%)
Laboratory tests	
ESR (mm/h), Me [25%–75%]	18 [6–28]
CRP (g/l), Me [25%–75%]	0.3 [0–6]
WBC (10 ⁹ /l), Me [25%–75%]	6.2 [4–8.7]
Hg (g/l), Me [25%–75%]	119 [97–128]
TA type (Moriwaki's classification)	
1	60 (33%)
2a	9 (5%)
2b	1 (0.5%)
3	3 (2%)
4	32 (17%)
5	78 (43%)

Note: Me — median; 25% — the lower quartile; 75% — the upper quartile.

bruits over the subclavian arteries or abdominal aorta, and a narrowed lumen or occlusion of the aorta or its major branches in the proximal upper and lower extremities not associated with atherosclerosis, fibromuscular dysplasia and spasm.

A reference dataset was compiled from Elibrary, a free Russian academic database of research works on biomedicine and natural sciences. We ran a search using *Takayasu's arteritis* and *TA* as keywords in order to obtain information about clinical and angiographic TA manifestations in Russian patients [1, 14, 19, 20].

Statistical analysis

The data were processed in Statistica 7.0 (Statsoft inc; USA). Categorical variables were analyzed using a chi-square test. All statistical tests applied were two-tailed; differences were considered significant at $p < 0.05$. The Kaplan–Meier method was used to analyze the factors affecting patients' survival. Between the groups, survival distributions were compared using the log-rank test.

RESULTS

Demographic data and clinical manifestations of the disease

At the time of examination, the most common clinical symptoms were malaise (47.5%), elevated blood pressure (53%), pulse asymmetry in the upper extremities (49%), pain, weakness or numbness in the upper extremities (49%), and headache (47%) (Table 2).

Erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and high-sensitivity CRP were elevated (Table 2). The most common pattern of arterial involvement was Moriwaki's type V (the aortic arch, its major branches, the abdominal aorta and/or renal arteries).

Pathological lesions were typically found in the subclavian, carotid and renal arteries. Stenosis was the most common pathology (Table 3).

Surgical interventions were performed on 66 (36%) patients (23 men and 43 women). Among the indications for surgery were: hemodynamically significant stenosis, occlusion or thrombosis of the involved vessels; hypertension caused by renal artery or subtotal aortic stenosis; symptoms of end-stage upper/lower limb ischemia. The surgical interventions performed can be broken down into two groups: endovascular procedures (stenting, bypass grafting, angioplasty) and open reconstructive surgeries. Besides, one patient with aneurysm and thrombosis of renal arteries had to undergo nephrectomy and kidney autotransplantation (Table 4).

In the majority of cases, medication therapy included glucocorticoids (96 patients; 52%) and antiplatelet agents, such as aspirin and dipyridamole (116 patients; 63%) (Table 5).

Since 1979, 31 patients have died, including 18 men and 13 women. The mean age at death was 38 years in women (Me [25%–75%] — 36 [32–44]) and 49 years in men (Me [25%–75%] — 50 [40–57]). On average, the time elapsed from the established diagnosis to the moment of death was 9.25 years (6.5 [3–16]) in men and 9 years (5 [3–10]) in women. Information about the causes of death was obtained from autopsy protocols ($n = 22$) and in conversations with the close relatives of the deceased ($n = 9$) (Table 6).

Data on the patients' survival is shown in Fig. 1. The 5-year survival rate was 92%; 10-year survival, 90%; and 15-year survival, 80%. (median: 34 [20–41]).

A total of 72 patients (27 men and 45 women) developed cardiovascular complications. On average, age at TA onset was Me [25%–75%] — 33 [26–43]. TA duration before the onset of a cardiovascular event was Me [25%–75%] — 10 [5–20]. Age at the onset of cardiovascular complications was 38 [30–49.5] (Table 7).

DISCUSSION

In spite of being considered a rare disease, Takayasu's arteritis is studied all over the world because it poses a significant social and economic burden: patients with TA develop serious cardiovascular complications causing disabilities and premature death in young people.

Although the arsenal of available diagnostic techniques is vast, the diagnosis of TA still remains a challenge due to the variability of its symptoms and the lack of knowledge of the disease demonstrated by public health practitioners. This leads to misdiagnosis during the initial examination or delays the correct diagnosis.

Our sample of the Ural residents was dominated by women: 139 (76%) of 183 participants were female. In other countries, the male to female ratio varies between 1 : 2.4 and 1 : 8 in Israel, 6.9 : 1 in Mexico and 8 : 1 in Japan [5–8]. In our study, the demographic characteristics, early clinical symptoms and the course of the disease were consistent with the previously published data indicating that TA strikes at young age [6–12].

Headache, hypertension (systolic blood pressure of 140 mmHg and/or diastolic blood pressure of 90 mmHg and above) were

Table 3. Arterial damage in the patients with TA (the present study)

Arteries involved	<i>n</i>
Subclavian	101 (55%)
Carotid	98 (53%)
Vertebral	16 (9%)
Axillary	11 (6%)
Brachial	12 (6.5%)
Pulmonary	6 (3%)
Coronary	31 (17%)
Celiac	42 (23%)
Superior mesenteric	45 (25%)
Renal	77 (42%)
Femoral	25 (14%)
Iliac	33 (18%)
Aortic arch	39 (21%)
Ascending thoracic aorta	15 (8%)
Descending thoracic aorta	9 (5%)
Abdominal aorta	52 (28.4%)
Lesion types	<i>n</i>
Stenosis	94 (51%)
Stenosis + occlusion	55 (30%)
Occlusion / Aneurysm	2 (1%)
Stenosis / aneurysm / occlusion	7 (4%)
Occlusion	8 (4%)
Stenosis + coarctation	3 (2%)
Stenosis + aneurysm	14 (7.6%)

Table 4. Surgical interventions performed in the patients with TA

Surgery type	Patients (n = 66)
Renal artery bypass with autogenous vein	4 (6%)
Carotid-subclavian bypass grafting	4 (6%)
Brachial artery bypass with autogenous vein	1 (1.5%)
Autogenous vein patch reconstruction of the brachial artery	1 (1.5%)
Renal artery stenting	13 (20%)
Descending thoracic aorto-bifemoral bypass grafting	3 (4.5%)
Thoracoabdominal bypass grafting	2 (3%)
Celiac trunk and superior mesenteric artery bypass grafting	3 (4.5%)
Abdominal aorta thrombectomy	2 (3%)
AFBG	7 (11%)
Renal artery angioplasty	11 (17%)
Carotid artery stenting	1 (1.5%)
Subclavian artery angioplasty	1 (1.5%)
Subclavian artery stenting	1 (1.5%)
Thrombectomy for thrombosis of a femoropopliteal bypass graft	1 (1.5%)
AFBG + SFA bypass grafting on the right side. Thrombosis of the aorto-femoral bypass graft/ graft thrombectomy in both branches. SFA and DFA angioplasty on the left, formation of a single ostium, profundoplasty on the right side	1 (1.5%)
Right renal artery dilatation	2 (3%)
Kidney autotransplantation	1 (1.5%)
Nephrectomy	4 (6%)
Renal artery thrombectomy	2 (3%)
BCA bypass grafting	2 (3%)
Left renal artery endarterectomy	1 (1.5%)
Subclavian-carotid anastomosis	2 (3%)
Celiac artery endarterectomy	1 (1.5%)
Brachial artery thrombectomy	1 (1.5%)
SFA thrombectomy on the right side	1 (1.5%)
Graft thrombectomy of the right branch	1 (1.5%)
Transfemoral amputation	2 (3%)
Abdominal revision for mesenteric thrombosis	1 (1.5%)
Infrarenal aorta bypass grafting	1 (1.5%)
Resection and end-to-end anastomosis of the small bowel following acute mesenteric thrombosis	1 (1.5%)
Resection of aneurysm in the brachiocephalic trunk	2 (3%)
Bifurcation alloplastic grafting of the aorta, carotid and brachiocephalic arteries	3 (4.5%)
Carotid artery thrombectomy	2 (3%)
Abdominal cavity and renal artery revision	2 (3%)
Resection of the right part of the large bowel and the terminal ileum + jejunotransverse anastomosis following acute mesenteric thrombosis	1 (1.5%)
Iliofemoral thrombectomy	1 (1.5%)
Implantation of arteriovenous graft in the left forearm	1 (1.5%)
Aorto-bicarotid bypass grafting	3 (4.5%)
Thoracoabdominal replacement following aortic coarctation	1 (1.5%)
Aneurysm resection in the abdominal aorta	1 (1.5%)
Aortic stenting following coarctation	1 (1.5%)
Resection of proximal anastomotic aneurysm in the subclavian artery	1 (1.5%)
Carotid-subclavian bypass grafting	1 (1.5%)
Autogenous vein repair of the subclavian artery	1 (1.5%)
Brachial artery dilatation	1 (1.5%)
Iliofemoral bypass grafting	1 (1.5%)
Coronary artery stenting	5 (7.5%)
AMCB	1 (1.5%)
DFA stenting on the right	1 (1.5%)
Linear iliofemoral bypass on the right side	1 (1.5%)

Note: AFBG — aorto-femoral bypass grafting; AMCB — aorto- and mammary coronary bypass; BCA — brachiocephalic artery; SFA — superficial femoral artery; DFA — deep femoral artery.

Table 5. Medication therapy received by the patients with Takayasu arteriitis

Medication therapy	Patients (n = 183)
Glucocorticoids	96 (52%)
Glucocorticoids + immunosuppressive agents (cyclophosphamide, azathioprine, hydroxychloroquine, methotrexate)	43 (23%)
No immunosuppressive therapy received or no exhaustive information about immunosuppressive therapy available in the medical records	40 (21%)
Genetically engineered drugs	0 (0%)
Antiplatelet agents	116 (63%)
Statins	27 (15%)
Nonsteroidal anti-inflammatory drugs	42 (23%)
Plasmapheresis	31 (17%)

Table 6. Causes of death in the patients with TA

Cause of death	Patients (n = 31)
Multiple cerebral infarctions resulting from progressive cerebral ischemia	1 (3%)
Cerebral infarction due to thrombosis of the internal carotid artery	1 (3%)
Bronchopneumonia	1 (3%)
Decompensated heart failure resulting from aortic regurgitation	2 (6%)
Massive bleeding due to anastomotic suture line failure following autogenous vein replacement of the renal artery	1 (3%)
Bronchopneumonia complicated by abscess after aorto-bicarotid bypass grafting, cerebral reperfusion injury and postoperative coma following implantation of a cardiac pacemaker	1 (3%)
Cerebral infarction + encephalopathy after cerebral reperfusion injury and postoperative cerebral coma after aorto-bicarotid bypass grafting	1 (3%)
Postoperative sepsis caused by suppurative mediastinitis after performing subclavian-carotid anastomosis	1 (3%)
Cerebral infarction due to thromboembolism following resection of brachiocephalic aneurysm	1 (3%)
Cerebral infarctions in the presence of progressive cerebral ischemia after bifurcation aorto-carotid bypass grafting	1 (3%)
Small bowel gangrene in the presence of intestinal ischemia after bifurcation aorto-femoral bypass grafting	1 (3%)
Postoperative peritonitis due to anastomotic suture line failure following small bowel resection for mesenteric thrombosis	2 (6%)
Intracerebral hemorrhage due to hypertension	2 (6%)
Pulmonary embolism caused by deep vein thrombosis of the lower extremities	1 (3%)
Pulmonary embolism caused by multiple arterial and venous thrombosis of the internal organs	1 (3%)
Cerebral infarction of the common carotid artery	1 (3%)
Massive bleeding due to the rupture of dissecting aortic arch aneurysm	1 (3%)
Massive bleeding due to the rupture of dissecting abdominal aortic aneurysm	2 (6%)
Acute renal failure caused by abdominal aorta thrombosis	1 (3%)
Peritonitis due to acute mesenteric thrombosis	1 (3%)
Myocardial infarction	2 (6%)
Hepatic cancer	1 (3%)
Myocardial insufficiency (ischemic stroke)	1 (3%)
Poisoning	2 (6%)
Unknown	2 (6%)

Note: PE — pulmonary embolism.

similarly frequent in all studies including our cohort of patients [9–14] (Table 8).

The most commonly involved vessels were the subclavian, carotid and renal arteries. Stenosis was the most common lesion type. The most prevalent (43%) TA type was type V (according to Moriwaki's classification). The carotid and subclavian arteries suffered the same types of lesions in all analyzed studies, including ours. Similar to other cohorts, the Ural sample demonstrated rare involvement of the vertebral, pulmonary and iliac arteries. Interestingly, damage to the ascending aorta and coronary arteries was very frequent in the Korean population [9, 15–18] (Table 9).

There are reports that the brachiocephalic artery is affected in 85% of Russian patients with TA, whereas damage to the renal artery is observed in 23% cases [19, 20] (Table 10).

Except for the Serbian study, all other research works indicate that type V of arterial involvement is the most frequent, while type III is the most rare. The Serbian sample is characterized by a high frequency of types I and IIa damage. Type IIb prevails in the Korean population. Type IV is relatively widespread in India and Brazil. Both genetic and environmental factors can contribute to the clinical manifestations of TA (Table 11) [15–17, 21–24].

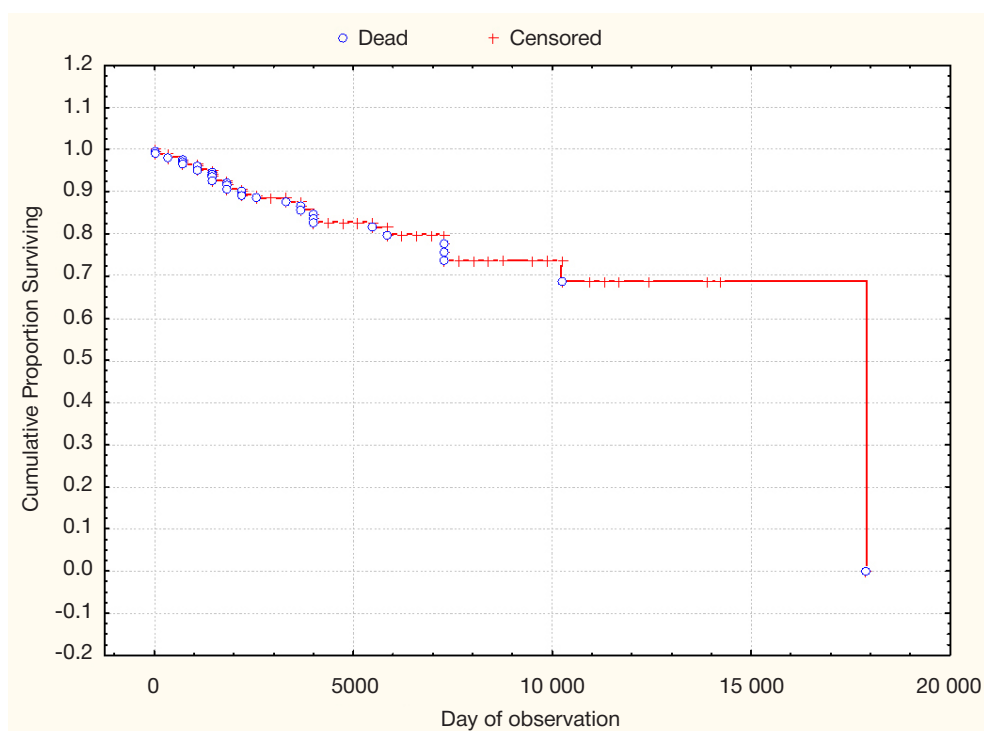


Fig. 1. Survival of the patients with TA

Table 7. Cardiovascular complications in patients with TA

	Patients with MACE (<i>n</i> = 72)
Myocardial infarction	14 (20%)
Ischemic stroke	24 (35%)
Transient ischemic attack	3 (3%)
Hemorrhagic stroke	4 (6%)
Thrombosis of the renal artery	5 (7%)
Thrombosis of the brachial artery	2 (3%)
Mesenteric thrombosis	4 (6%)
Thrombosis of the radial artery	1 (1.4%)
Thrombosis of the brachiocephalic trunk	1 (1.4%)
Thrombosis of the dorsalis pedis artery	2 (3%)
Thrombosis of the axillary artery	1 (1.4%)
Thrombosis of the infrarenal aorta	1 (1.4%)
Thrombosis of the abdominal artery	2 (3%)
Thrombosis of the carotid artery	3 (3%)
Thrombosis of the subclavian artery	3 (3%)
Celiac thrombosis	1 (1.4%)
Aneurysm rupture in the thoracic aorta	2 (3%)
Aneurysm rupture in the abdominal aorta	1 (1.4%)
Thrombosis of the aorto-femoral bypass graft	6 (8%)
PE	3 (3%)
Thrombosis of the jugular vein	2 (3%)
Cerebral venous sinus thrombosis	1 (1.4%)
Thrombosis of the sural veins	1 (1.4%)
Thrombosis of the small saphenous vein	1 (1.4%)
Thrombophlebitis of the tibial veins	1 (1.4%)
Thrombophlebitis of the aorto-venous bypass graft of the renal artery	1 (1.4%)

Table 8. Clinical symptoms of TA in different geographical regions

	This study (n = 183), %	Italy (n = 67), % [9]	India (n = 106), % [10]	Brazil (n = 73), % [11]	South Africa (n = 272), % [12]	Japan (n = 84), % [13]	Russia (n = 215), % [14]
Malaise	47.5	n/a	n/a	n/a	10	n/a	50
Weight loss	24	12	9.4	28	n/a	5	9
Fever	35.5	39	16	26	10	20	n/a
Headache	47	33	47	45	n/a	n/a	50
Dizziness	22	27	n/a	29	n/a	n/a	n/a
Pain, weakness, numbness of the upper extremities	49	52	59	58	12	54	50
Hypertension (systolic pressure of 140 mmHg and diastolic pressure of 90 mmHg)	53	46	52	36	77	52	68
Pulse asymmetry in the upper extremities	49	73	59	85	12	54	50
Blood pressure difference > 10 mmHg	20	72	59	85	12	54	50

Table 9. Arteries affected by the pathology in patients with TA residing in different geographical regions

Arteries	This study (n = 183), %	China (n = 411), % [15]	Korea (n = 20), % [16]	USA (n = 126), % [17]	Italy (n = 104), % [9]	France (n = 82), % [18]
Subclavian	53	79.8	67.1	6.3	65.6	68.3
Carotid	55	79.1	72.1	50.9	44.3	59.8
Vertebral	9	28.7	n/a	18.5	13	28.0
Pulmonary	3	68.9	13.4	33.3	n/a	n/a
Coronary	17	35.7	63.3	22.2	n/a	n/a
Mesenteric	25	29.7	22.8	24.7	31.6	n/a
Renal	42	48.9	32.2	18.7	34.4	14.6
Iliac	18	27.2	13.3	13.5	19.7	18.3
Ascending aorta	8	9.5	47.8	9.1	n/a	n/a

Table 10. Damage to the arteries in patients with TA representing the Russian population

Arteries	This study (n = 183)	Central Russian regions [19]	North European Russia [20]
Subclavian	55	85%	n/a
Carotid	53		n/a
Vertebral	9		n/a
Pulmonary	3	n/a	n/a
Coronary	17	n/a	n/a
Mesenteric	25	n/a	n/a
Renal	42	n/a	23%
Iliac	18	n/a	n/a
Ascending aorta	5	n/a	n/a

Table 11. Angiographic characteristics of TA in different regions across the world

	This study	China [15]	Korea [16]	USA[17]	India [21]	Mexico [22]	Serbia [23]	Brazil [24]
Type I, %	33	22.1	11.1	20	6.9	19	50	11.9
Type IIa, %	5	3.9	8.6	6	1	3	19	6
Type IIb, %	0,5	3.9	14.1	7	5.9	4	0	1
Type III, %	2	2.9	4.0	5	2.9	4	0	9
Type IV, %	17	6.3	7.6	5	28.4	2	0	27
Type V, %	43	60.8	54.5	57	54.9	69	31	18

In our cohort of patients, survival rates were similar to those reported in previous publications [17, 25–27]. However, there are reports of different 5- and 10-year survival rates (69 and 36%, respectively) [28]. In the Arab population, the 5-year survival rate is as low as 50% [29]. The record-breaking 100% survival rate is reported in Japan [30].

A national TA registry is needed to estimate the prevalence of the disease in different ethnic groups. It will provide valuable information about TA presentation and clinical course necessary to establish the timely diagnosis. The actual prevalence of TA can be higher than suggested in the literature. Timely diagnosis and treatment will help to improve patient outcomes.

The main limitation of the present study is its retrospective design. Besides, the study was conducted in inpatients only. In different time periods, the diagnostic procedures used in the patients were different, which also complicates data interpretation.

CONCLUSIONS

The clinical presentations of TA are diverse. Timely diagnosis can be a challenge when a patient presenting with general inflammation has a healthy pulse pattern, develops collateral circulation and demonstrates no specific symptoms of arterial damage. Patients with TA should be warned against a high risk of cardiovascular complications and advised to monitor and control their blood pressure, lipid counts and blood coagulability. Hemodynamically significant stenosis, occlusion or thrombosis of damaged arteries can be corrected surgically, contributing to antihypertensive therapy, helping to eliminate the symptoms of ischemia and prevent new vascular complications. The majority of patients in our cohort had Moriwaki's type V of arterial damage. High survival rates can be explained by a young age and a capacity to develop collateral circulation.

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