DOI 10.12851/EESJ201503C01ART04

Irina E. Borodina,
postgraduate;

Lubov A. Shardina,
MD, Professor,
Ural State Hospital № 1, Ural State Medical University;

Ten Year Study of Manifestations in Patients with Nonspecific Aortoarteritis in Urals

Keywords: *Non-specific aortoarteritis, angiography.*

Annotation: *The article presents the results of a 10 year period study from 2005 to 2015 of clinical, laboratory and angiographic manifestations of Takayasu arteritis in russian patients in Urals.*

Non-specific aortoarteritis (NSAA ,Takayasu's arteritis) is a systemic inflammatory disease of aorta and its branches, which leads to stenosis and ischemia of surrounding tissues. The disease is prevalent worldwide it is more common in developing Afro- Asian countries. Etiology of the Takayasu's disease is unknown. Syndrome of the systemic inflammatory reaction with the clinical manifestations of the effects of lesions of the aorta and its branches at different anatomical levels (symptom: coarctation, abdominal ischemia, coronary, renovascular hypertension, lesions of the pulmonary artery) are criteria of this systematic process (6).

Takayasu's arteritis (TA) is similar to other forms of vasculitis, including giant cell arteritis which typically affects older individuals (1). The first case of Takayasu's arteritis was described in 1908 by Japanese ophthalmologist MikitoTakayasu at the Annual Meeting of the Japan Ophthalmology Society. Takayasu described a peculiar "wreathlike" appearance of the blood vessels in the back of the eye (retina) (8).

It is generally considered a disease of young adults with a peak onset in second and third decades of life and a striking predilection for females. The predisposing and etiological factors of TA have still to be clearly elucidated. An autoimmune basis, influenced by genetic and environmental factors, is strongly suggested; the resulting inflammation is largely a cell-mediated immune response. (5).

In TA, biopsy specimens are seldom available and hence morbid anatomic features are based on autopsy findings or segments excised during bypass surgery. On histology, the lesions can be active, chronic, or healed. Though TA is a panarteritis, the initial site of inflammation is around the vasa vasora and at the medio-adventitial junction (4,8) .The iam of

a study is to identify the main clinical, laboratory and angiographic features of Takayasu arteritis in Russian patients over a 10 year period from 2005 to 2015.

Materials and methods:

This study included 66 patients over a 10 year period from 2005 to 2015 at Sverdlovsk State Hospital № 1, Ekaterinburg. The inclusion criterion were the existence of a least three of the American College of Rheumatology (ACR) criteria (1):

- Age at disease onset < 40 years (Development of symptoms or findings related to Takayasu arteritis at age 40 year).
- Claudication of extremities (Development and worsening of fatigue and discomfort in muscles of 1 or more extremity while in use, especially the upper extremities).
- Decreased brachial artery pulse (Decreased pulsation of 1 or both brachial arteries). 4. BP difference >10 mm Hg (Difference of >10 mm Hg in systolic blood pressure between arms)
- Bruit over subclavian arteries or aorta (Bruit audible on auscultation over 1 or both subclavian arteries or abdominal aorta)
- Arteriogram abnormality (Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or similar causes; changes usually focal or segmental).

Activity of the disease, according to NIH guideline (8-10) (2,3)

- Systemic signs and symptoms such as fever and arthralgia
- Increased ESR or CRP levels
- Signs of vascular insufficiency (pulses of variable intensity in the extremities or pulselessness, limb claudication)
- New lesion in serial vascular imaging in previously normal vessels

Exclusion criteria included:

- Other connective tissue disorders
- Autoimmune diseases
- Infectious diseases
- liver diseases
- thyroid diseases
- metabolic syndrome
- diabet
- nephrotic syndrome
- alcoholism
- drug-addition
- mental disorders
- oncology

Non-specific aortoarteritis angiographic classification is (2):

- Type I - Branches of the aortic arch (arch cervicobrachial)
- Type II- thoracoabdominal aorta
- Type III - abdominal aorta, renal arteries ,descending aorta
- Type IV – combination of types I, II, III

Laboratory tests included ESR, white blood cell (WBC), rheumatoid factor (RE). Imaging studies included: ECG, chest X ray, echocardiography, angiography.

Results

There were 66 patients , 46 of them (69 %) were females and 20 (30 %) were males. The mean age of females was 44 years. The mean age was 47 years. The time interval between the beginning of symptoms and diagnosis of non-specific aortoarteritis was 3 months to 5 years.

Clinical manifestations in females.

There were 46 females. The mean age was 44. The mean age at the time of diagnosis was 24,8. The most frequent symptoms at the time of diagnosis were fatigue in 16 patients (34,7 %), headache in 25 females (54,3 %), dizziness in 19 patients (41,3 %), hypertension in 23 females (50%).

The most frequent symptoms at the moment of observation were headache in 15 females (32,6 %), dizziness in 12 patients (26 %), hypertension in 20 females (43,4 %) (table 2).

In laboratory tests, ESR range was 19-56 mm/h with a median of 37 mm/h. 19 of females(41,3 %) had ESR more than 25 mm/h at the moment of diagnosis. High level of CRP more than 5 mg/L was in 7 females (15,2 %). 15 females (32,5 %) had leukocytosis with a WBC count of more than 10000. Rheumatoid factor lever was positive in 3 females (6,5 %). Hemoglobin range was 85 -130 gm/dl with median of 104 gm/dl.

According to angiographic findings 10 females (21,7 %) had type 1, 5 females (10,8 %) had type 2, 12 females (26 %) had type 3, 19 females (41,3 %) had type 4.Regarding vascular lesions, the most common lesion in angiography was stenosis in 42 females (91,3 %), artery occlusion had 11 females (23,9%), coartation was founded in 1 female (2.1 %), aneurysm in 4 females (8,6 %). The most common involved left subclavian artery (15 females, 32,6 %), left carotid artery (11 females, 23,9 %), right renal artery (17 females, 36,9 %), left external iliac artery (10 females, 21,7 %).

According to echocardiographic findings left ventricular hypertrophy was founded in 3 females (6,5 %), aortic regurgitation had 7 females (15,2 %), ejection fraction lower than 50 % was founded in 3 females (6,5 %), diastolic dysfunction of left ventricular was founded in 7 females (15,2 %).

According to complications 5 of females (10,8 %) had strokes and 1 of females (2,1 %) had infarction myocardial.

Clinical manifestations in males.

There were 20 males. The mean age was 47. The mean age at the time of diagnosis was 21.2. The most frequent symptoms at the time of diagnosis were dizziness in 8 patients (40 %), hypertension in 10 males (50 %), limb claudication in 7 patients (35 %).

The most frequent symptoms at the moment of observation were fatigue in 5 males (25 %), headache in 8 patients (40 %), hypertension in 15 males (75 %) and limb claudication in 8 males (40 %).

In laboratory tests, ESR range was 22-60 mm/h with a median of 39 mm/h. 8 of males (40%) had ESR more than 20 mm/h at the moment of diagnosis. High level of CRP more than 5 mg/L was in 2 males (10 %). 9 females (45 %) had leukocytosis with a WBC count of more than 10000. Rheumatoid factor lever was negative. Hemoglobin range was 108 -132 gm/dl with median of 112 gm/dl.

According to angiographic findings 4 males (20 %) had type 1, 2 males (10 %) had had type 2, 5 males (25 %) had type 3, 9 patients (45 %) had had type 4.Regarding vascular lesions, the most common lesion in angiography was stenosis in 15 males (75 %), artery occlusion had 6 females (30 %), coartation was not founded, aneurysm in 2 males (10 %).The most common involved right renal artery (7 males, 35 %), left external iliac artery (6 males,30 %), left external iliac artery (7 patients, 35 %), right femoral artery (6 patients,30 %).

According to echocardiographic findings left ventricular hypertrophy was founded in 6 males (30 %), aortic regurgitation had 3 males (15 %), ejection fraction lower than 50 % was founded in 2 females (10 %), diastolic dysfunction of left ventricular was founded in 5 females (25 %).

According to complications 3 of males (25 %) had strokes and 3 of males (25%) had infarction myocardial.

Conclusion

66 patients were observed. There were 46 females and 20 males. The mean age of females was 43 years; the mean age of males was 39 years. Headache and hypertention were the most common presentation in females, dizziness and hypertention were the most common presentation in males. The angiography manifestations were classified as type 1 (10 females/ 4 males), type 2 (5 females/2 males), type 3 (12 females/5 males), type 4 (19 females/9 males).

Table 1.Clinical manifestations at the time of diagnosis

	Females (46)	Males (20)
Fatigue	16 (34,7 %)	4 (20 %)
Fever	8 (17,3 %)	5 (25 %)
Weight loss	11(23,9 %)	4 (20 %)
Myalgia	7 (15,2 %)	4 (20 %)
Arthralgia	4 (8,6 %)	2 (10 %)

Visualproblems	8 (17,3 %)	3 (15 %)
Memory problems	7 (15,2 %)	5 (25 %)
Headache	25 (54,3 %)	5 (25 %)
Dizziness	19 (41,3 %)	8 (40 %)
Chest pain	5(10,8 %)	2 (10 %)
Hypertension High blood pressure (more than 140/90 mm	23(50 %)	10 (50 %)
Abdominal pain	10 (2,17 %)	3 (15 %)
Claudication	11 (23,9 %)	7 (35 %)
Weak pulses or pulseless upper extremities	10 (2,17 %)	2 (10 %)
Difference in blood pressure in upper extremities	8 (17,3 %)	2 (10 %)
Weak pulses or pulseless lower extremities	11 (23,9 %)	6 (30 %)

Table 2. Clinical manifestations at the moment of observation

	Females (46)	Males (20)
Fatigue	8 (17,3 %)	5 (25 %)
Fever	2 (4,3 %)	2 (10 %)
Weight loss	6 (13 %)	3 (15 %)
Myalgia	7 (15,2 %)	2 (10 %)
Arthralgia	2 (4,3 %)	1 (5 %)
Visualproblems	8 (17,3 %)	4 (20 %)
Headache	15 (32,6 %)	8 (40 %)
Dizziness	12 (26 %)	2 (10 %)
Tachycardia	9 (19,5 %)	3 (15 %)
Hypertension High blood pressure (more than 140/90 mm	20 (4,3 %)	15 (75 %)
Abdominal pain	13 (28,2 %)	5 (25 %)
Claudication	11 (23,9 %)	8 (40 %)
Weak pulses or pulseless upper extremities	10 (2,17 %)	3 (15 %)
Weak pulses or pulseless lower extremities	9 (19,5 %)	6 (30 %)

Table 2.Angiographic manifestations in females and males

Types	Females (46)	Males (20)
Type 1	10 (21,7 %)	4 (20 %)
Type 2	5 (10,8 %)	2 (10 %)
Type 3	12 (26 %)	5 (25 %)
Type 4	19 (41,3 %)	9 (45 %)

The most common clinical, laboratory and angiographic findings were hypertension, increased ESR, lower hemoglobin and stenosis. Making the diagnosis of Takayasu's arteritis can be extremely difficult. Combination of these findings, which we founded may be helpful for physicians to define diagnosis. Our study will be useful in future research related to Takayasu disease in Urals.

References:

1. *American College of Physicians (ACP). Medical Knowledge Self-Assessment Program (MKSAP-15): Rheumatology. "Systemic Vasculitis", 2009, ACP; 65-67.*
2. *Kerr GS. Takayasus arteritis: Rheum. Dis. North Am., 1995, Vol. 214; 1041-1058.*
3. *Kerr GS, Hallahan CW, Giordano J. et al. Takayasu arteritis: Ann. Intern. Med., 1994, Vol.12011; 919-929.*
4. *Kinare SG, Gandhi MA, Deshpande JR. Non-specific aortoarteritis (Pathology and Radiology) Mumbai: Quest Publications; 1998.*
5. *Kothari SS. Aetiopathogenesis of Takayasu's arteritis and BCG vaccination: The missing link. Med Hypotheses. 1995; 45; 227–30.*
6. *Mel'nik MV, Shilov AM, Uvarovskaia BV, Men'shikova IV, Pak IuV. Early diagnosis of Takayasu's disease: Clinical case report, 2014, Nov-Dec; 59(6): 52-6.*
7. *NumanoF. Differences in clinical presentation and outcome in different countries for Takayasus arteritis: Curr.Opin.Rheumatol, Vol.91; 12-15.*
8. *Seko Y. Takayasu arteritis: insights into immunopathology: Jpn. Heart J. 2000, Vol.411; 15-26*
9. *Takayasu. A case with peculiar changes of the central retinal vessels. ActaSocietatisophthalmologicaeJaponicae, Tokyo, 1908, 12; 554.*