



Takayasu Arteritis

Alternative Names

Aortic Arch Syndrome
Young Female Arteritis
Pulseless Disease
Reverse Coarction

WHO International Classification of Diseases

Diseases of the circulatory system

OMIM Number

207600

Mode of Inheritance

? Autosomal recessive vs. autoimmune

Description

Takayasu's Arteritis (TA) is a rare, chronic, inflammatory disease that predominantly involves the aorta and its branches (mostly thoracic and abdominal). Patients might be either asymptomatic or present with severe neurological symptoms. Fibroproliferation occurs in aorta, great vessels, pulmonary arteries, and renal arteries that results in segmental stenosis, occlusion, dilatation, and aneurysmal formation in these vessels. TA is the only form of aortitis that causes stenosis and occlusion of the aorta. "Pulseless disease" is referred to TA because there is difficulty in detecting peripheral pulses which may be the cause of vascular narrowing. In approximately 90% of cases, TA appears in patients younger than 30 years with a female: male ratio of 8:1. About six persons per 1000 are affected worldwide. However, most TA cases are reported in Asia.

TA is divided into early active phase (inflammatory) and chronic late phase. The late-phase is further subdivided into four types depending on the involved site. Type I (classic pulseless type) involves the brachiocephalic trunk, carotid arteries, and subclavian arteries; type II is a combination of type I and III; type III (atypical coarctation type) involves the

thoracic and abdominal aortas distal to the arch and its major branches; and type IV (dilated type) involves extensive dilatation of the length of the aorta and its major branches. However, the most common type is type III (65% of patients). Corticosteroids have been the treatment of choice until now.

Molecular Genetics

The cause of Takayasu's arteritis (TA) is not known, but there are several hypotheses that suggest the etiology of the disease. One hypothesis proposes that the origin of TA is infectious or autoimmune. Genetic susceptibility to TA has been extensively studied. It is found that there are heterogeneous population data considering the associations of human lymphocyte antigen (HLA) with TA.

Epidemiology in the Arab World

Bahrain

Al Jishi et al. (2005) described, for the first time, a 41-year-old married woman with a combination of Takayasu arteritis (TA) and primary antiphospholipid antibodies (aPL) syndrome who underwent carotid stenting. In the first observation, she had a normal ECG. After eight years, early vasculitis was detected on biopsy from erythematous skin lesion. Also, her upper limb pulses were weak and a weak flow through the right radial was noticed by ultrasound. One year later, she was admitted to neurology ward with history of dysarthria of acute onset. She was suffering from bronchial asthma for 15 years. On examination, she was obese, and her speech was slurred. She had upper motor neuron left facial palsy, and left carotid bruit. MRI and MRA of the neck and brain and arch aortogram displayed severe stenosis of the left common carotid artery (CCA), and occlusion of the right brachiocephalic artery and of left subclavian artery. X-ray, ECG, echocardiogram, and



cardiac catheterization showed heavily calcified aortic root, arch and descending aorta; 30% osteal calcification of the left main coronary; and first degree of heart block. Human lymphocyte antigen (HLA) analysis was obtained showing that the patient had HLA-A2 and A9 which were susceptibility alleles for TA in Arabs. She had single nucleotide polymorphism (SNP) double homozygosity for methylenetetrahydrofolate reductase (MTHFR); C677T (T/T genotype) and A1298C (C/C genotype). Serum homocysteine was elevated and that might be attributed to C677T SNP. It was thought that MTHFR 1298C/C genotype would be an independent risk factor for ischemic stroke. In addition, she carried the D/D genotype of the insertion- deletion mutation of Angiotensin converting enzyme and was E3/E3 genotype carrier of apolipoprotein E. The patient met two major diagnostic criteria for TA which were pulselessness and occluded left subclavian artery in addition to some minor criteria (hypertension, aortic regurgitation, left mid common carotid lesion). She underwent CCA stenting, however she did not receive steroids that would prevent restenosis because she could not tolerate it. She developed acute episode of aphasia four months after the stenting.

Iraq

Baumgartner et al. (2005) described a 14-year-old Iraqi girl with a 3-year history of fever, fatigue, malaise and diffuse pain. Ultrasound and magnetic resonance angiography showed marked thickening of the aortic wall, dilatation of the aortic arch, and decreased luminal diameters of the abdominal aorta and both subclavian arteries, consistent with TA. Ascending and descending aortic elastic properties such as distensibility and stiffness index were markedly reduced compared to a group of healthy controls (n=39): ascending aortic distensibility was 20 kPa(-1) x 10(-3) versus 63+/-23 kPa(-1) x 10(-3) in controls, and

the ascending aortic stiffness index 9.6 versus 3.5+/-1.3 in controls.

Lebanon

Atallah et al. (1998) reported on a series of 42 renal artery stenosis treated with percutaneous transluminal angioplasty (PTA) from January 1988 to June 1996. Among the 33 patients described 22 presented an atheromatous pathology, seven a fibromuscular dysplasia. One patient had a Takayasu arteritis, and 3 others a stenosis of the renal graft artery.

Tunisia

Ben Zineb et al. (1992) reported a case of a pregnant woman with Takayasu's arteritis and Crohn's disease. The course of pregnancy was uneventful. Ben Zineb et al. (1992) suggested that these two inflammatory diseases may be immunologically related.

References

- Al Jishi A, Krishnan PR, Almawi WY. Takayasu arteritis with high titre of antiphospholipid antibodies and MTHFR Polymorphism. *J Thromb Thrombolysis*. 2005; 20(1):47-50.
- Atallah N, Smayra T, Slaba S, Menassa L. [Percutaneous renal angioplasty. Experience of the Radiodiagnostic Service of the Hotel-Dieu of France] *J Med Liban*. 1998; 46(5):244-50.
- Baumgartner D, Sailer-Hock M, Baumgartner C, Trieb T, Maurer H, Schirmer M, Zimmerhackl LB, Stein JI. Reduced aortic elastic properties in a child with Takayasu arteritis: case report and literature review. *Eur J Pediatr*. 2005; 164(11):685-90.
- Ben Zineb N, Zine S, Bellasfar M, Mesaad MJ, Sfar R. [Association of Takayasu arteritis, Crohn disease and pregnancy] *Rev Fr Gynecol Obstet*. 1992; 87(12):591-3.

Contributors

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