

Takayasu arteritis in a young female

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Abstract

Background: Takayasu arteritis is a granulomatous vasculitis which mainly involves the large arterial vessels. The disease is rare and it is more common in females aged between 15-30 years old. In this paper, we report a case of takayasu arteritis in Babol, north of Iran.

Case Presentation: A 22 - year old female was admitted to the Department of Infectious Diseases of Rouhani Teaching Hospital due to anemia, high erythrocyte sedimentation rate (ESR), low grade fever and weight loss in the early summer of 2011. Her problems started four years ago and was hospitalized in another hospital and after full investigation, no diagnosis was found. Physical exam showed a vascular problem, then MRI angiography was done and showed an involvement of right carotid, abdominal aorta and right iliac arteries and the diagnosis of Takayasu Arthritis was confirmed. Prednisolone was administered, her fever subsided and ESR, CRP and hemoglobin were normalized.

Conclusion: Takayasu arteritis should be considered in the differential diagnosis of patient with protracted low grade fever, anemia and dramatically elevated ESR in young female in order to prevent late complications of the disease.

Key words: Takayasu, Treatment, Diagnosis, Elevated ESR, Young female.

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Takayasu's arteritis or aortic arch syndrome is a large vessel granulomatous vasculitis affecting often young or middle-aged women of Asian descent (1, 2). It has a worldwide distribution with the greatest prevalence in Asians (3-5). The disease mainly affects the aorta and its main branches, as well as the pulmonary arteries. Females are affected about 8-9 times more than males. The symptoms and signs of the disease begin between 15 and 30 years of age. Due to obstruction of the main branches of the aorta, including the brachiocephalic artery, and the left subclavian artery, takayasu's arteritis can present a decrease or absence of upper extremities pulses and for the same reason it is also called pulseless disease (6, 7).

Some patients develop an initial inflammatory phase characterized with malaise, fever, night sweats, weight loss, arthralgia, and fatigue. Initial inflammatory phase is often followed by the symptoms of vascular insufficiency manifesting arms or leg claudication, hypertension due to renal artery stenosis, and neurological manifestations like lightheadedness and syncope. The neurological symptoms of the disease vary depending on the distribution and degree of the blood vessel involvement (8-10). The one rare but important feature of the takayasu's arteritis is ocular involvement in the form of visual field defects, visual loss, or retinal hemorrhage (11, 12).

The most important laboratory findings are anemia and marked elevation of the ESR or C- reactive protein (1, 2). The gold standard of diagnosis is vascular study by arterial angiography (DSA), Magnetic resonance angiography (MRA), computed tomography angiography (CTA). Inflammation, granuloma, and fibrosis causes stenosis (93%), occlusion (57%), dilatation (16%), aneurysm (7%) in subclavian artery (93%), common carotid artery (58%), renal artery (38%), vertebral artery (35%) (13-18).

Recently researchers have shown the presence of serum antiendothelial cell antibodies increased soluble E-selectin and thrombomodulin in the serum of the patients. The value of these tests and also PET scan in diagnosis or follow-up is still uncertain (19, 20).

Case presentation

A 21- year old female was admitted to the Department of Infectious Diseases of Rouhani Teaching Hospital due to anemia, high erythrocyte sedimentation rate (ESR), and low grade fever and weight loss in the early summer of 2011. Her problems started four years ago. She was also admitted in a hospital in Tehran but after full investigations no diagnosis was found. She frequently visited several physicians for her treatment during these years without any improvement. In physical examination: temperature was 38°C, BP in left arm 110/80 mmHg, right arm 100/80 mmHg, left leg 130/80 mmHg and right leg 140/80 mmHg. She had pale conjunctiva, heart and pulmonary sound, and abdominal exam were normal, except few lymph nodes in bilateral inguinal area, peripheral pulses were symmetric but bruite in auscultation of abdominal aorta, both carotid arteries, right subclavian artery was heard. Chest X-ray, echocardiography, abdominal sonography were normal.

WBC; 15500/ml, (poly=80%, lym=20%), ESR=130 mm/h, CRP=4+

MRangiography showed stenosis of abdominal aorta, below the renal arteries, and right carotid, right iliac artery and color Doppler study showed thickness of both carotid, subclavian, axillary arterial walls and increase of PSV of these vessels. By these findings, the diagnosis of takayasu arteritis was confirmed and the patient was transferred to the Department of Rheumatology for further evaluation and treatment (figure 1-3). Treatment with high dose glucocorticoids and methotrexate and aspirin was started.



Figure 1. Stenosis of abdominal aorta



Figure 2. Stenosis of right and left carotid arteries

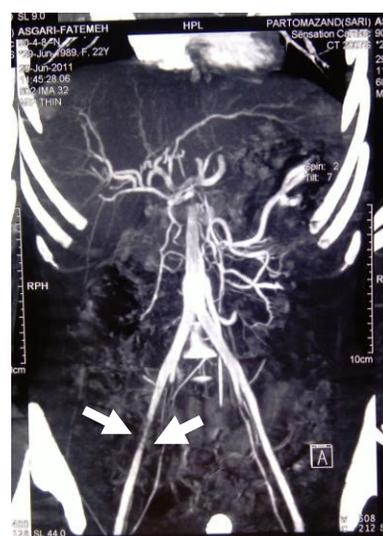


Figure 3. Stenosis of a right iliac artery

Discussion

In this article, we present a rare case of takayasu arteritis after approximately four years delay. This disease is a granulomatous vasculitis that originally affects the large arteries. The inflammatory processes cause inflammatory injury that results narrowing, occlusion, or dilation of involved arteries in varying degrees (11). The most clinical symptoms and signs of the disease include arthralgias or myalgias occurring in about one-half of cases. Articular symptoms can be transient or continuous over several months or longer. The pulmonary arteries are involved pathologically in up to 50 percent of cases (14). Involvement of the carotid and vertebral arteries causes decreased cerebral blood flow, leading to vertigo, syncope, headaches, convulsions, and dementia. Visual impairment is a late manifestation and is due to cerebral ischemia (8). Our patient

described arthralgia, myalgia, vertigo and syncope which were transient and it seems that they are related to her disease. The most key features of this disease are pulselessness or asymmetric pulses between the arms or significant discrepancy between blood pressure of upper extremities. Our patient did not have this sign and we believe that it was one of the reasons of late diagnosis in this patient. The first case of Takayasu's arteritis was described in 1908 by a Japanese ophthalmologist and he described a peculiar "wreathlike" appearance of the blood vessels in retina (21). It is now known that the blood vessel malformations that occur in the retina are an angiogenic response to the arterial narrowings in the neck, and other large vessels (9). In conclusion, Takayasu arteritis should be considered in the differential diagnosis of every young female with protracted low grade fever, anemia and dramatically elevated ESR in order to prevent late complications of the disease.

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