Case report

A 54-year-old male patient with amaurosis fugax

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ABSTRACT

Takayasu arteritis is an idiopathic chronic large vessel vasculitis. It is a rare chronic disease of the aorta and its branches, and is mostly seen in children and young women. The clinical picture includes non-specific systemic symptoms along with those related to the problematic artery. However, patients can sometimes be asymptomatic at the time of diagnosis. The most common symptoms include upper extremity claudication, systemic hypertension, pain around the carotid artery, dizziness, and vision problems. Diagnosis is based on clinical criteria and the golden rule in identifying arterial lesions in Takayasu arteritis is angiography. Herein we report a 54-year-old male patient with amaurozis fugax, who was later diagnosed as Takayasu arteritis

Key Words: Amaurosıs Fugax, Takayasu Arteritis, vasculitis, vision loss, panarteritis

Takayasu arteritis (TA) is a form of idiopathic chronic large vessel vasculitis involving the aorta and its main branches, the pulmonary arteries, and the coronary tree[1]. TA was first described by Mikito Takayasu in 1908 and predominantly affects women of reproductive age[2]. Clinical presentation varies according to the location of the arterial lesions[2].

CASE

A 54-year-old male patient with 7-8 short-lived episodes of vision loss (Amaurosıs fugax) in two years referred for a consultation. Detailed anamnesis revealed effort-independent and atypical chest pain in the retrosternal area in the last six months. History of the patient revealed 8 years of hypertension, smoking until recently, and an aortovertebral bypass surgery due to interrupted aortic arch branching 28 years ago. Following this surgery, the patient had not receive any medical treatment or had complaints until 2 years ago. His physical examination revealed a significant difference between the blood pressure measurements of the right (150/100 mmHg) and left (110/75 mmHg) arms. Additionally, the left arm radial pulse could not be taken. His physical examination revealed no further pathological findings; nor did his biochemical examination, complete blood count, sedimentation, telecardiography, electrocardiography and echocardiography results. As a further examination, the patient also received vertebral artery angiography (including aortography). The angiography showed that the left subclavian artery was totally blocked except for a short segment (Figure 1). The left vertebral artery received retrograde flow during right vertebral artery injection. In the left vertebral artery proximal, left thyreocervicalis turuncus arteries were seen (subclavian steal) (Figure 2). Also, the left anterior serebral artery and its branches received flow from the right carotid artery. In accordance with the 1990 diagnosis criteria of

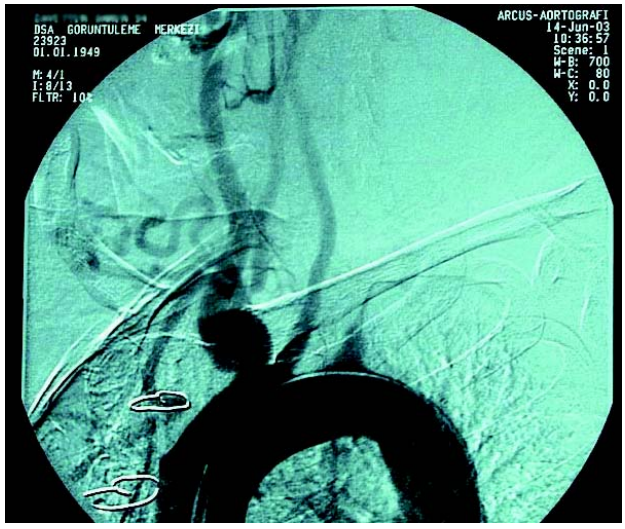

Figure 1. Angiography showed the left subclavian artery was totally blocked

American College of Rheumatology, the patient was diagnosed with TA (Table I). The findings

leading to this diagnosis included complaints before the age of 40, absent left radial pulse, a significant difference between the blood pressure of the two arms, interrupted aortic arch branching, and blockage in the left subclavian artery. Since TA may also be seen in coronary arteries[1], coronary angiography was performed on the patient to account for the chest pain. The results came back normal.

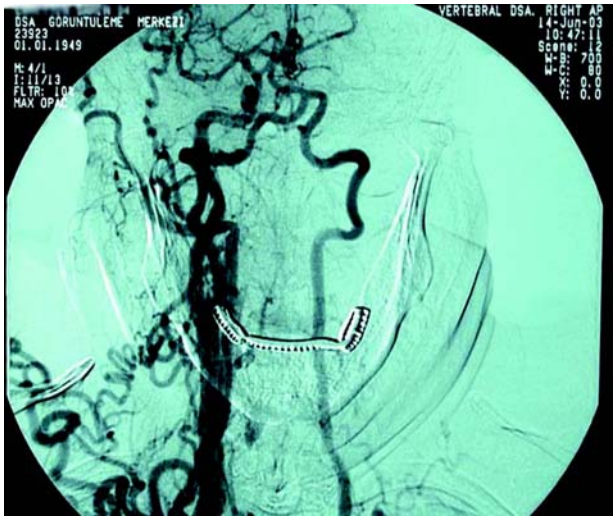

Figure 2. The left vertebral artery received retrograde flow during right vertebral artery injection. In the left vertebral artery proximal, left thyreocervicalis turuncus arteries were seen (subclavian steal)

Table I. Takayasu's arteritis: The American College of Rheumatology's 1990 diagnostic criteria[1]

- Patient below 40 years of age at initial diagnosis
- Claudication of extremities
- Reduced brachial pulse
- Difference in blood pressure between right and left arms >10 mmHg
- Bruit above subclavian artery, aorta or both
- Arteriographic appearance

At least three criteria must be present for diagnosis.

DISCUSSION

The annual rate of TA is 2.6/1 million worldwide[2]. Most patients are between 10 and 30 years of age. It is a rare chronic disease of the aorta and its branches, and is mostly seen in children and young women. It affects females four times as much as males[2]. Apart from the aorta and its main branches, it may also be detected in the pulmonary and coronary arteries[1].

The histological lesions of TA are the focal panarteritis picture associated with the stages of the disease[3]. Histologicaly, lymphocytes, plasma cells, histiocytes and multinuclear giant cells, and polymorphonuclear cells infiltration can be seen[2]. In the further stages of the disease, the normal structure of the vascular system containing internal and external elastic lamina is deformed. This process results in intimal wall thickness, emergence of scars, and eventually thrombosis[2]. In addition aneurism develops especially in the weakened vein wall[3]. The clinical picture includes non-specific systemic symptoms along with those related to the problematic artery. However, patients can sometimes be asymptomatic at the time of diagnosis[3]. The most common symptoms include upper extremity claudication, hypertension, pain around the carotid artery, dizziness, and vision problems[4]. 43% of the cases also display non-specific symptoms such as fever, fatigue, weight loss, and night sweats. As the disease worsens, cardiac symptoms often become more visible with the contribution of aortic insufficiency, coronary artery disease and hypertension[4].

Physical examination often reveals absent or reduced pulse in one or more primary veins such as the radial or bracial arteries, and 10 mmHg or more difference in the blood pressure of the two arms[1]. At present, there is no appropriate serological test for the diagnosis of TA. Laboratory tests are non-specific[5]. Anemia and mild leucocytosis may accompany the disease. 72% of the patients display high levels of sedimentation, which is normalized in 56% of the patients during the remission stage[2]. In the active stage, the patient may display low albumin and high globulin[2]. Diagnosis is based on clinical criteria and the golden rule in identifying arterial lesions in TA is angiography[3].

The first stage treatment for TA is immune suppression. With glucocorticoid treatment, 40 to 60% remission may be observed[1]. Initially, the dose for glucocorticoid treatment is 60 mg daily or 1 mg/kilogram until they stabilize. The dose is then reduced gradually[2]. Approximately 40% of the patients resist steroids[1]. In patients resistant to corticosteroids, treatment must be accompanyied by such cytotoxic agents as cyclosphosphamide, methotrexat, mycophenylate mofetil, and cyclosphorin[2]. Lesions that respond to the initial steroid treatment may eventually turn into stenosis, and vascular bypass surgery may be needed. A combination of medical treatments and agressive surgery has reduced mortality rates; however, around 20% of the patients resist all treatment[2].

TA mostly seen in woman. Male patients are rare especially in older ages. Symptoms may disappear with surgery or medical therapy but because of vascular complications, late ischemic symptoms can be developed without reactivation of TA after many years. These symptoms may be confused with atherosclerosis.

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