Case Report

TAKAYASU'S ARTERITIS

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ABSTRACT

Takayasu's arteritis is a large vessel vasculitis whose histopathologic findings are indistinguishable. Takayasu's arteritis is seldom reported because of being a rare disease. The incidence of Takayasu's arteritis is 5 to 8 times higher in Japan, India, and Africa than in the United States. This disease occurs mostly in young women, preferentially affecting the aorta and its major branches, and causes stenotic and ectatic changes. The best method for its evaluation is angiography and managements are medical and surgical.

Keywords: Aorta,Subclavian artery, Carotid artery, Angiography, Hypertension

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INTRODUCTION

In 1908, Takayasu, a Japanese ophthalmologist, reported retinal arteriovenus shunts in a *wreath-like* distribution around the optic disc and micro aneurysms of the retinal vessels in a 19-year-old Japanese woman.

In an ensuing discussion, Onishi related similar findings in a patient with pulseless, cool upper extremities, the etiology of the described ocular and vascular findings was at that time obuscure but has since become known as Takayasu's arteritis (TA).

It is now recognized that, although the arterial involvement may be confined to the aortic arch as in Takayasu's original patient, involvement of multiple aortic segments or entire aorta and its major branches, and pulmonary artery can occur.

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Arterial pathology varies from a fulminant accute inflammatory process to extensive post inflammatory mural fibrosis. Infectious, hereditary, and autoimmune etiologies have all been suggested, yet a definitive demonstration of a causal relationship has been elusive.

CASE REPORT

Among 507 patients referred to angiography ward due to vascular disorders in upper limbs and carotids, such as stenosis, aneurysms and embolisms, three cases of Takayasu's arteritis were diagnosed. It is also possible to employ other modalities, for instance colour Doppler and MRI in diagnosis. However angiography is most commonly known as a kind of "gold standard" for diagnosis of this disorder. Though a great number of the above-mentioned patients had previously undergone other modalities, they were sent to angiography ward for final diagnosis.

Case 1: This was a 37-year–old female patient from Makoo, whose illness started initially by general weakness, occasional fever and headache followed by decrease of vision. She had consulted the regional physicians and in spite of long term treatment she had obtained no benefit. Her illness particularly her general

weakness and headache further aggravated in intensity gradually. She was referred to dispensary of Imam Khomeini Hospital for further investigations. Physical examination of patient revealed weakened radial and carotid pulses, and because of that the patient was transferred to the Cardiac Center of Shahid Madani Hospital. The patient was primarily hospitalized in internal ward of that center but for the decreasing pulses amplitude that was on the way of vanishing was presented to the angiography department of Imam Khomeini Hospital. In angiograms, as shown in (Figure 1a-b), occlusion of the common carotid and subclavian arteries is noticed bilaterally and brain perfusion is achieved only by vertebral arteries. Blood supply for the extremities occurred by extensively weakened collaterls. Great vessels, especially renal arteries appeared normal in angiography through abdominal aorta (Figure 1c).

Case 2: A 30-year-old lady from Mianeh was admitted to Imam Khomeini Medical Center, due to mild pain and decreased strength of the right upper limb and was hospitalized in internal medicine ward for management. Several investigations resulted in the possible diagnosis of Takayasu disease in this setting, and the patient was referred for angiography, which showed complete occlusion of the right subclavian artery. Blood supply to the right upper extremity was yielded by collateral arteries (Figure 2). This patient is actually under the supervision and follow-up of the internist colleagues, as she has refused to be operated.

Case 3: This is a female patient of 35 years of age from Tabriz who was admitted and hospitalized in internal ward of the Imam Khomeini Hospital because of headache, coldness and weakness of the left upper limb and vision disorders of the left eye. After intensive investigations the patient was referred to the angiography department with possible diagnosis of Takayasu disease. There was evidence of complete obstruction of the common carotid and subclavian arteries on the left side in

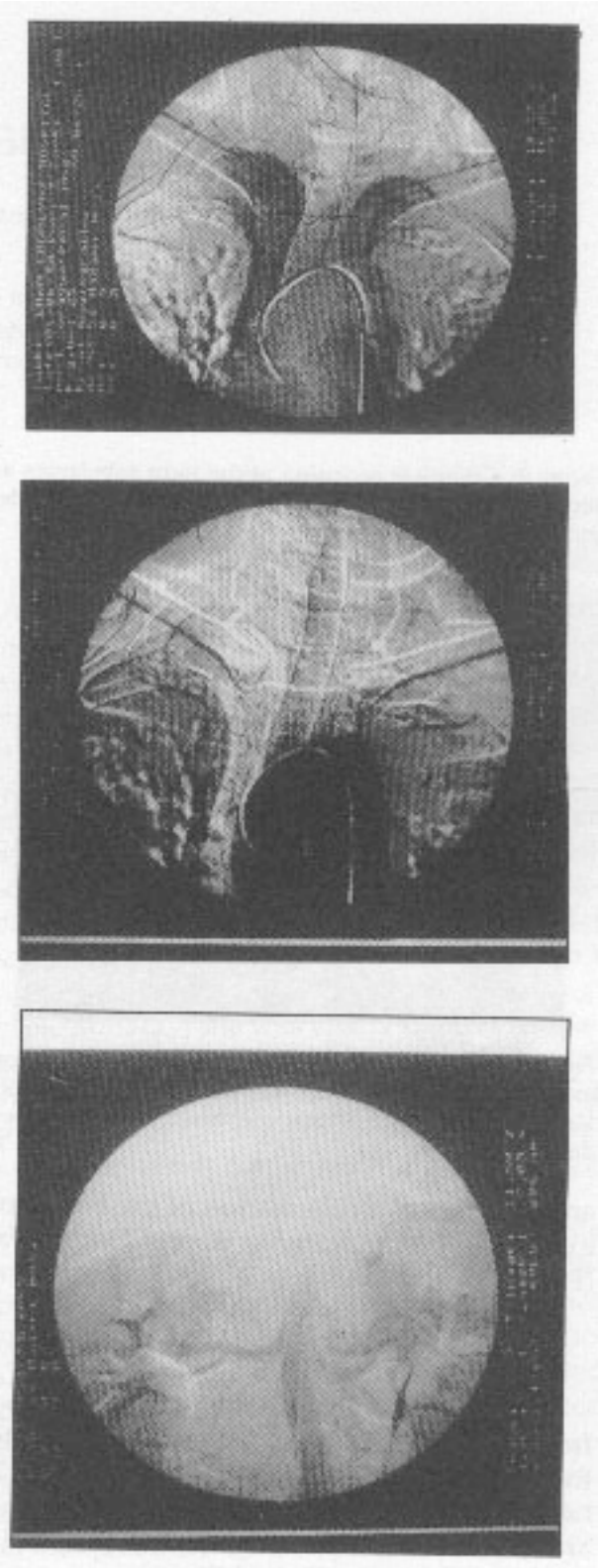

Figure 1: (a-b-c) Occlusion of the common carotid and subclavian arteries is noticed bilaterally and brain perfusion is achieved only by vertebral arteries.

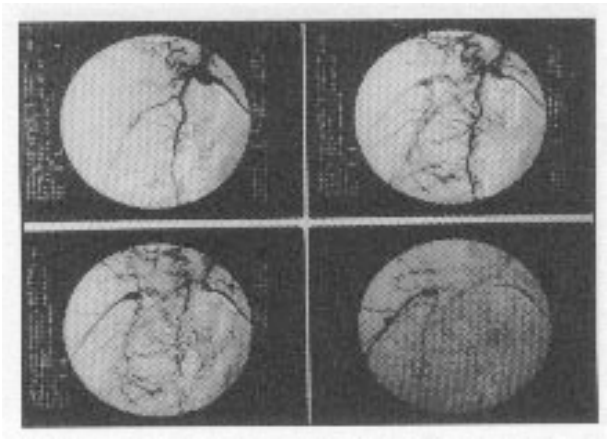

Figure 2: Complete occlusion of the right subclavian artery. Blood supply to the right upper extremity was yielded by collateral arteries.

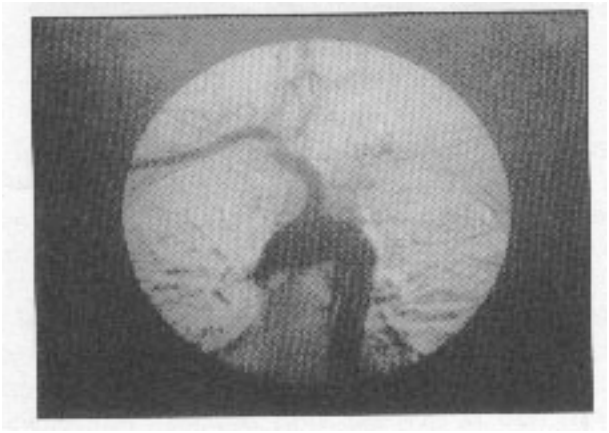

Figure 3: Complete obstruction of the common carotid and subclavian arteries on the left side in angiography. Vascularisation of the left upper extremity was imperfectly done by collaterals.

angiography. Vascularisation of the left upper extremity was imperfectly done by collaterals (Figure 3).

DISCUSSION

Takayasu's arteritis is a rare disease. A study from Olmstead County, Minnesota estimated the incidence of Takayasu's arteritis to be 2.6 cases per year. The incidence of Takayasu's arteritis is 5 to 8 times higher in Japan, India, and Africa than in United Stated.

Patients with Takayasu's arteritis may initially experience systemic manifestations such as fever, malasie, weight loss, arthralgia, and myalgia. Later, patients develop symptoms of arterial occlusion (e.g., dizziness, headache, claudication, exertional dyspenia, or chest pain). Examination at this stage almost always reveals absent or reduced peripheral pulses or bruits. Hypertension is common and may be missed in patients with bilateral subclavian disease because of falsely low blood pressure readings in the arms.[1]

The inflammatory process results in segmental stenoses, occlusions, and aneurismal degeneration of the aorta and proximal arterial tree. This results in a variety of clinical presentation that may include renovascular hypertension, renal failure, stroke, ocular ischemia, extremity ischemia, aortic insufficiency, or aortic aneurysm formation.[2,3]

The benefits of revascularization for these conditions are well recognized, but the unique pathogenesis and uncertain natural history of TA have tempered enthusiasm for a surgical approach to this entity. Thus, surgical therapy has been traditionally reserved for severe symptomatic manifestations of arterial occlusive disease refractory to high-dose corticosteroids or other medicinal therapies.

There was one perioperative death, a 19-year-old woman who had elective resection of a true aneurysm of the left ICA (histology demonstrates Takayasu's arteritis).[4,5]

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