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Case Report

Takayasu's Arteritis in a Libyan Female

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Abstract

Takayasu's arteritis (TA) is a large-vessel vasculitis that involves the aorta and its major branches. Renal arteries are frequently involved, usually with renovascular hypertension. The prevalence of TA in Arabs is low. A study of the epidemiological and clinical features of TA in Arabs included 197 identified patients between 1995 and 2012 and none of them was Libyan. We report a 61-year-old Libyan woman in whom TA manifested with hypokalemia and arterial hypertension. Previous ultrasound showed renal size asymmetry raised the possibility of renal artery stenosis. The diagnosis of TA was confirmed by magnetic resonance angiography, which showed a thickened abdominal aortic wall, occlusions of the left renal artery and left common iliac artery, stenosis of the right common iliac artery, and stenosis of both subclavian arteries. TA is rarely encountered in Arabs. However, the disease must be considered in patients who present with renovascular hypertension, in a context of other autoimmune disorders.

Keywords: Hypokalemia, hypothyroidism, insidious, Libya, renal artery stenosis, Takayasu's arteritis

Introduction

Takayasu's arteritis (TA) is a chronic vasculitis, characterized by chronic granulomatous inflammation of the vessel wall of large- and medium-sized arteries with a predilection for the aorta and its major branches as well as the proximal portions of pulmonary, coronary, and renal arteries.[1] It can lead to progressive stenosis, occlusion, or aneurismal transformation, with subsequent significant morbidity and mortality.[1]

The pathogenesis of TA is still unknown; an interplay of genetics and autoimmune factors has been suggested.[2] It has been described in association with autoimmune disorders, such as systemic lupus erythematosus, rheumatoid arthritis, systemic sclerosis, ankylosing spondylitis, granulomatosis with polyangiitis, Crohn's disease, and hyperthyroidism.[2-8] Although the disease has a worldwide distribution, and it is more prevalent among Eastern and Southeast Asian populations, the prevalence in Japan was 40 cases/million population compared to 4.7–8.0 per million population in the rest of the globe.[2] The clinical manifestation of TA varies depending on the site and severity of vascular involvement.

A study of the epidemiological and clinical features of TA in Arab countries included 197 patients identified between 1995 and 2012. Among Arabs, the renal artery was involved in 20%–50% of the patients and hypertension was present in approximately one-third. None of these patients was from Libya. We report a case of TA diagnosed during medical evaluation of incidentally discovered hypokalemia in a Libyan patient with hypothyroidism.

Case Report

A 61-year-old Libyan female presented with a medical history of hypothyroidism on L-Thyroxine replacement since 2012, vitiligo since 1980, and chronic sinusitis. She has been on regular follow-up at the endocrine clinic. The results of her routine laboratory studies revealed a low serum potassium level. She denied any constitutional symptoms. There was no prior history of arthralgias, myalgias, or any other symptoms, suggestive connective tissue or atherosclerotic peripheral vascular disease. No symptoms of loss of muscle strength, visual impairment, syncopal episodes, vertigo, or abdominal pain after eating were observed. No suggestive symptoms of renal or gastrointestinal Potassium loss were noted. She

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denied any use of diuretics, laxative, or herbs. Drug history included intake of levothyroxine 75 µg and simvastatin 20 mg once daily. Her average blood pressure (BP) recording at the outpatient clinic was 130/80 mmHg, and she denied taking any antihypertensive medication. She had a history of surgery for carpal tunnel syndrome in 2012. A discrepancy of kidney size was noted on the ultrasound abdomen in 2009. The patient reported that there was difficulty in measuring BP in her right arm. On examination, she looked well, clinically euthyroid, not anemic or jaundiced. Thinning of the scalp hair and vitiligo patches were present [Figure 1]. Her weight was 72.5 kg, height was 151 cm tall, and body mass index was 31.8 kg/m^2. Pulses of the left brachial and radial arteries were not palpable, her pulse rate was 82 beats/min, and BP in the right arm was 140/90 mmHg and in the left arm was 110/70 mmHg. The rest of the cardiovascular and respiratory examination did not reveal any abnormalities. The abdomen was soft, lax, with no organomegaly and no renal bruits. Laboratory parameters are presented in Table 1. Thyroid and liver function tests were normal. Magnetic resonance angiography revealed stenosis of both subclavian arteries, thickened abdominal aortic wall, occlusions of the left renal artery and left common iliac artery, and stenosis of right common iliac artery [Figure 2]. She was referred to the rheumatology clinic for further evaluation and management.

Discussion

TA was named in honor of Japanese Ophthalmologist Mikito Takayasu (1859–1938), who first reported a case of the disease in 1905.[9] TA is a large-vessel vasculitis that involves the aorta and its major branches. It occurs most commonly in Asia particularly in young females.[2] In 1990, the American College of Rheumatology proposed six criteria for the diagnosis of TA [Table 2]. Three out of six criteria are needed for the diagnosis, with a sensitivity of 90.5% and a specificity of 97.8%.[10] Angiographic classification[11] defined six types of TA based on the anatomic distribution of vascular involvement is summarized in Table 3. Angiographic classification correlates with clinical manifestations and prognosis but cannot differentiate active from burned-out lesions. According to the arterial angiography study, our patient belongs to Type V, with the involvement of ascending aorta, aortic arch, and its branches and abdominal aorta and/or renal arteries.

Table 1: Some relevant laboratory parameters of the patient and the laboratory reference ranges

Variables	Results	Reference values
White blood cell count	9400/mm^3	4000-11,000
Hb	15.2 g/dl	12-16
Platelet count	265000/mm^3	150,000-400,000
Erythrocyte sedimentation rate (1st h)	40 mm/h	<20
Serum C-reactive protein	1.25mg/dl	<6
Antinuclear antibodies index	0.2	<1.0
Fasting plasma glucose	121mg/dl	<126
HbA1c (%)	5.9	<5.7
Total serum cholesterol	206 mg/dl	<200
Serum triglycerides	130mg/dl	<150
Serum HDL-C	37mg/dl	>40
Serum sodium	136 mmol/L	136-146
Serum potassium	3.1 mmol/L	3.5-4.5
Chloride	102 mmol/L	98-107
e-GFR	68.2 ml/min/1.73 m^2	-
Serum renin	7.7 ng/l	2.6-28
Aldosterone	17.4 ng/l	2.2-35
Renin:aldosterone ratio	22.5	<19
Serum TSH	2.34 mU/ml	0.27-4.2
Serum T4	107 nmol/L	66-187
Serum AST	26.8 U/L	<40
Serum ALT	45.6 U/L	<41
Serum ALP	118 U/L	50-136

AST: Aspartate transaminase, ALT: Alanine transaminase, ALP: Alkaline phosphatase, e-GFR: Estimated glomerular filtration rate, HDL-C: High-density cholesterol, HbA1c: Hemoglobin A1C, TSH: Thyroid stimulating hormone

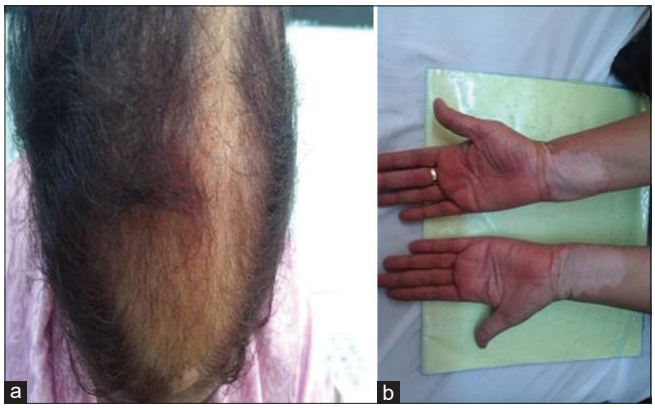

Figure 1: Clinical features suggestive of associated autoimmune diseases. (a) Thinning of scalp hair, (b) vitiligo patches

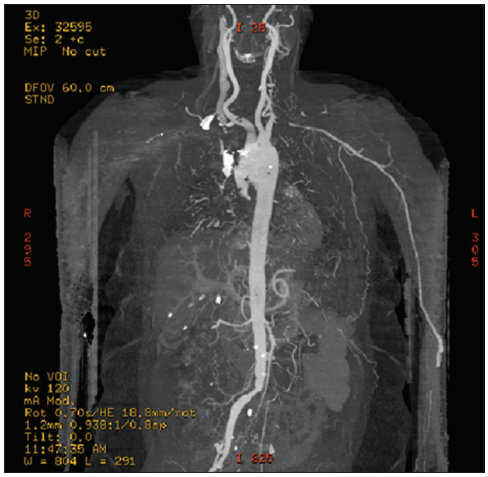

Figure 2: Magnetic resonance angiography of the aorta, demonstrating stenosis of both subclavian arteries, thickened abdominal aortic wall, occlusions of the left renal artery and left common iliac artery, and stenosis of right common iliac artery

Table 2: The six American College of Rheumatology criteria for the diagnosis of Takayasu's arteritis

1. Onset before 40 years of age
2. Claudication of the extremities
3. A decrease in the brachial pulse in one or both arms
4. A difference of 10 mmHg or more in BP measured in both arms
5. Audible bruit on auscultation of the aorta or subclavian artery
6. Narrowing at the aorta or its primary branches on the arteriogram

Three out of the six criteria are needed for the diagnosis to be made. Arend *et al*. BP: Blood pressure

Table 3: The angiographic classification of Takayasu's arteritis defining six types based on the anatomic distribution of vascular involvement

Type	Involved vasculature
Type I	Branches from the aortic arch
Type IIa	Ascending aorta, aortic arch, and its branches
Type IIb	Ascending aorta, aortic arch and its branches, thoracic descending aorta
Type III	Thoracic descending aorta, abdominal aorta, and/or renal arteries
Type IV	Abdominal aorta and/or renal arteries
Type V	Entire aorta and its branches (i.e., combined features of types IIb and IV)

Moriwaki *et al*.

Although the disease typically begins in the second or third decades of life, it is not uncommon in the extremes of age. Soto *et al*. from Mexico reported 9% of their patients at diagnosis as aged >40 years; 10.7% of patients studied by Vanoli *et al*. were >50 years; 4.9% of patients in a French study were aged >60 years.[12-14] Time lag to diagnosis is approximately 15 months. A delay in diagnosis of 2–11 years is seen in the West.[2] In our patient, the previous ultrasound report of a discrepancy of renal size pointed to a disease onset beyond 10 years, with a chronic, insidious course.

A recent study of the epidemiological and clinical features of TA among Arab populations showed female predominance. The age average at onset in most series was 31.5 years, and the mean delay in diagnosis was 3.5 years (ranged 1.5–4.2 years).[15,16] A long delay in the diagnosis, was in part due to the low awareness of a relatively rare disease.

Patients can present in an early stage of disease activity with constitutional features, such as fever, nocturnal sweats, weight loss, headache, and malaise; with or without symptoms caused by vascular stenosis, occlusion, or aneurysms; or may diagnose later in the "burnout" or chronic stage.[15]

Clinical manifestations of the disease vary depending on the sites and severity of vascular lesions. In our patient, the incidental finding of hypokalemia triggered re-assessing the patient history, examination, and investigation. Presence of hypokalemia with a discrepancy in renal size, in an older person with a history of hypertension, hypothyroidism, and hyperlipidemia, raises the probability of atherosclerosis as the cause of renal artery stenosis (RAS). The insidious onset of the disease, the absence of the initial acute inflammatory phase of TA, and the rarity of the condition in Libyans may have contributed to the delay in the diagnosis.

Conclusions

TA is a rare condition. Only a few cases were reported in the international literature from Libya.[17] The disease was diagnosed in the burned-out stage during evaluation of an incidental finding of hypokalemia secondary to RAS. Delays in diagnosis can be reduced by carefully searching for unequal or absent upper extremity pulses and by listening for renal bruits in hypertensive patients. The disease must be considered in patients who present with renovascular hypertension, in a context of other autoimmune disorders.

Declaration of patient consent

The author certifies that she obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

Compliance with ethical principles

No prior ethical approval is required for single case reports. However, the patient provided consent for publication, as stated above.

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