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Takayasu Arteritis: A Diagnostic Approach

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ABSTRACT

Takayasu arteritis (TA) is a systemic, inflammatory large-vessel vasculitis of unknown etiology that most commonly affects women of childbearing age. It involves the aorta and its major branches. The main abnormalities found in this disease are vascular ischemic signs/symptoms including claudication, carotid bruit, weak distal pulsation. Hereby we report a case a 25-year-old unmarried female complained a recurrent left arm pain, worsening when she worked as a physician. Formerly, she had experienced intermittent bilateral leg pain for four years with strange sound coming from inside her neck. She was hospitalized half a year ago due to acute limb ischemic and she was diagnosed with TA after the imaging showed multiple stenosis in large arteries including bilateral neck area, shoulder area, abdominal aorta, and right renal artery. The inferior right extremity biopsy showed the histologist result of erythema nodosum appearance. C Reactive Protein (CRP) Quantitative is 19.8, and Erythrocyte Sediment Rate (ESR) is 53. The others laboratory data to exclude differential diagnosis are unremarkable. This report demonstrates the importance of diagnostic approach in rare case of Takayasu's arteritis patients.

1. Introduction

Takayasu's arteritis is a chronic granulomatous vasculitis affecting large arteries: primarily the aorta and its main branches.¹ The data on the epidemiology of TA is limited, it could be due to the rarity of the disease. Although the disease has a worldwide distribution, it is generally thought to be much more common among Asian populations. The incidences of TA were estimated to be 1–2 per million in Japan and 2.2 per million in Kuwait. Recent epidemiologic studies suggest that TA is being increasingly recognized in Europe with reported incidence estimates varying from 0.4 to 1.5 per million. The highest ever prevalence of TA at 40 per million was estimated in Japan and the lowest ever one at 0.9 per million in US.²

Vasculitis syndromes are classified, according to the size of the affected vessels, into large-vessel, medium-vessel, and small-vessel vasculitis. Large-sized vessel

vasculitis, ie, vasculitis occurring in the aorta and its major branches to the extremities, the head and the neck, includes Takayasu arteritis.³

Takayasu's arteritis may occur either as a primary process or secondary to other associated conditions. Secondary vasculitis may be associated with connective tissue disease, infection, malignancy, drugs or other factors.

The American College of Rheumatology (ACR) includes arteriogram abnormalities in the diagnostic criteria of the disease. Angiography is the gold standard for evaluation of vascular lesions. Angiography allows a topographic classification which correlates anatomic involvement, clinical manifestations and prognosis: in 1994 the Takayasu Conference in Tokyo proposed an angiographic classification that allows to distinguish

different subgroups of patients with six patterns of involvement with the appendage of a “C” or “P” to denote coronary or pulmonary involvement, respectively.⁴

It is important to identify active diseases since it requires immunosuppressive treatment, but it is unclear whether immunosuppression is effective in the late stage of the disease when a risk-benefit balance must be made.

Case Presentation

A 25-year-old unmarried female complained a recurrent left arm pain, worsening when she worked as a physician. Formerly, she had experienced intermittent bilateral leg pain for four years with strange sound coming from inside her neck. She was hospitalized half a year ago due to acute limb ischemic (there was no flow of leg arteries) and there were blood pressure (right arm): 140-182/104-132 mmHg, (left arm): unmeasurable, (right leg): 149/98, (left leg): 165/107. Upper extremities: radial artery pulsation left < right. Lower extremities: dorsalis pedis artery pulsation right < left; tibialis posterior artery, poplitea artery, and femoral artery were normal from physical examination. She was diagnosed with TA after the imaging showed multiple stenosis in large arteries including bilateral neck area, shoulder area, abdominal aorta, and right renal artery (**figure 1, 2**). She had undergone laboratory examination and the results found were leucocytosis with positive ANA 1/100, anti ds-DNA 1.0, non-reactive of HBsAg and anti HCV, ACA IgG 2.3, IgM 10.4, PT: 10.6/10.9, aPTT: 30.9/32.1, fibrinogen: 272.4, D dimer: 0.2, electrolyte within normal limit, albumin: 3.8, random glucose: 103, aspartat transaminase: 46.8, alanin transaminase: 43.1, ureum: 40.9, creatinine: 1.14, Beta2 GP IgG :2.4 Beta 2 GP IgM: 5.3, Quantitative CRP: 19.8, Rheumatoid Factor: <10.4, level of C3 : 126, C4: 32.30, HDL/LDL/total cholesterol/triglycerida: 37/72/128/167, ESR: 53. The inferior right extremity biopsy showed the histology result of erythema nodosum appearance. Neurosonology laboratory result (carotid and vertebral arteries examination): occlusion

of left vertebra artery and stenosis of bilateral common carotid artery and right vertebra artery.

2. DISCUSSION

Our patient with complain of a recurrent left arm pain which worsened with activity which consistent with limb claudication. Findings from physical examination also revealed evidence of arterial involvement. Site of affected vessel should be the entry point for making diagnostic approach in vasculitis suspected patient. In accordance with patient complain and physical examination, the vascular involvement in this patient is large vessel (**Table 1**).⁵

With the involvement of large arteries of the extremities, diagnosis of TA should be considered therefore, we proceed with imaging examination. CT findings also consistent with TA. When a patient suspected as vasculitis, we have to ensure the vasculitis due to primary or secondary by excluding infection, malignancy, collagen disease and other factors. The laboratory data to exclude secondary vasculitis or differential diagnosis are unremarkable (**figure 3**).⁶

The patient's inferior right extremity biopsy showed erythema nodosum appearance as the histology result. Cutaneous manifestations had been observed in up to 28% of patients with Takayasu's arteritis. They include pyoderma gangrenosum (mainly in Asian patients), erythema nodosum (predominantly in Caucasian patients), as well as erythema induratum.⁷

According to American College of Rheumatology (ACR) Classification Criteria of Takayasu arteritis (**Table 2**), the patient was diagnosed as TA because of age at disease onset <40 years, claudication of extremities, decreased brachial artery pulse, blood pressure difference >10 mmHg and arteriogram abnormality.⁸

Using angiographic classification of Takayasu's arteritis (**Table 3**), the patient was diagnosed as type V TA based on imaging that showed multiple stenosis in large arteries including bilateral neck area, shoulder area, abdominal aorta, and right renal artery. Therefore patient should be considered to advance therapy.⁴

Patients were categorized having active disease in accordance to features of vascular ischaemia or inflammation (claudication, diminished or absent pulses, vascular pain, asymmetric blood pressure), lab result that showed increasing Quantitative CRP and

ESR, and typical angiographic finding.⁹ Finally, the patient confirmed as type V vessel involvement based on angiographic classification of Takayasu arteritis, with active disease activity.

Figure 1. Stenosis a. inominate, bilateral carotiscommunis arteries, bilateral subclavian arteries.

Figure 2. Stenosis abdominal aorta, infrarenal level. Stenosis right renal artery with contracted right (staghorn calculi)

Table 1. Typical clinical manifestation of large, medium, and small-vessel involvement by vasculitis

Large	Medium	Small
Constitutional symptoms: fever, weight loss, malaise, arthralgia/arthritis		
Limb claudication	Cutaneous nodules	Purpura
Asymmetric blood pressure	Ulcers	Vesiculobullous lesions
Absence of pulses	Livedo reticularis	Urticaria
Bruits	Digital gangrene	Glomerulonephritis
Aortic dilation	Mononeuritis multiplex	Alveolar hemorrhage
	Microaneurysms	Cutaneous extravascular
		Necrotizing granulomas
		Splinter hemorrhages
		Uveitis
		Episcleritis
		Scleritis

Figure 3. Approach to the diagnosis of vasculitis syndrome. **Table 2.** The 1990 American College of Rheumatology Classification Criteria for Takayasu arteritis

Criteria	Definition
Age at disease onset < 40 years	Development of symptoms or findings related to TA at age < 40 years
Claudication of extremities	Development and worsening of fatigue and discomfort in muscles of 1 or more extremity while in use, especially the upper extremities
Decrease brachial artery pulse	Decreased pulsation of 1 or both brachial arteries
Blood pressure difference > 10 mmHg	Difference of > 10 mmHg in systolic blood pressure between arms
Bruit over subclavian arteries or aorta	Bruit audible on auscultation over 1 or both subclavian arteries or abdominal aorta

Arteriogram abnormality

Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or similar causes: changes usually focal or segmental

A patient shall be classified as TA if at least 3 of these 6 criteria are present

Table 3. Angiographic classification of Takayasu's arteritis.

Type	Vessel involvement
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	Combined features of types IIb and IV

3. CONCLUSION

The diagnostic approach of vasculitis should consider vessel vasculitis involvement, type of angiographic classification and disease activity.

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