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Case Report

CASE REPORT

Pivotal role of cardiac magnetic resonance imaging in a new case of Takayasu arteritis

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Abstract

Takayasu arteritis (TA), also known as the pulseless disease, is a form of vasculitis of unknown cause that chiefly affects the aorta and its major branches, most frequently in young women. The earliest detectable abnormality in TA is a thickening of the vessel wall, but diffuse aortic wall calcification is very rare and is a late manifestation. Besides, the involvement of the coronary arteries is not a common finding in TA and frequently involves the right coronary artery (RCA). Multi-modality imaging has a fundamental role in the diagnosis of vasculitis and its complications. In this report, we want to present an unusual case with TA, diffuse aortic wall calcification and left main coronary artery ostial lesion, which is a rare combination.

INTRODUCTION

Takayasu Arteritis (TA) is a type of vasculitis affecting aorta and its major branches, thought to result from an autoimmune process that targets large elastic-containing arteries. TA affects women eight times more frequently than men. Chronic inflammation of the vessel walls leads to aneurysm formation, stenosis or thrombosis. Cardiac involvement occurs eventually in nearly one-third of patients. Myocarditis also occurs in TA and causes potentially reversible congestive heart failure (CHF). Conventional angiography is invasive and provides the least sensitive method for visualizing wall thickness. Ultrasonography, and to a lesser degree, computed tomography (CT) can detect this early vessel wall thickening [1]. Magnetic resonance angiography (MRA) can identify vessel wall thickening, stenosis, as well as aneurysm formation. Additionally, multi-parametric cardiac magnetic resonance (CMR) imaging assists in determining cardiac involvement, including myocarditis. In this article, we want to present a new case of TA with atypical features including diffuse aortic wall calcification, left main (LM) coronary artery ostial thickening, as well as myocarditis.

CASE REPORT

A 37-year-old man, from Mazandaran, Iran referred to our hospital with a 4-month history of palpitation and dyspnea (NYHA functional class II). The patient had no medical history of diabetes, hypertension, dyslipidemia or coronary artery disease. Also, no history of smoking or alcohol use, as well as shortness of breath and chest pain and angina symptoms during exercise was detected. However, he had a history of sudden cardiac death in his brother at the age of 37 years old, with no evaluation. Moreover, he had no evidence of arthralgia, fever, headache or dermatologic manifestation at the time of presentation. On clinical examination, his blood pressure was 120/80 mm Hg in both upper limbs, pulses were easily palpable, and there were no bruits. A systolic murmur with an intensity of II/VI heard

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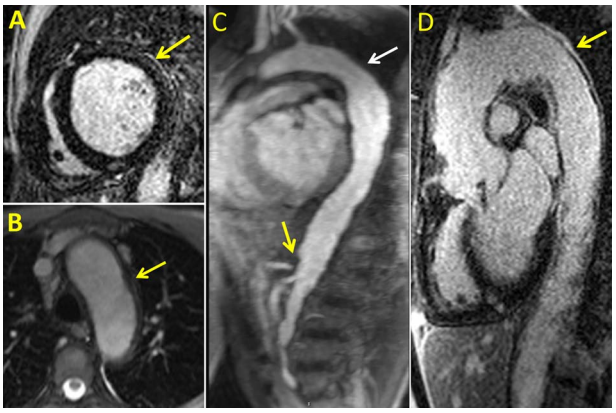

Figure 1: CT and angiography in TA. (A) CT angiography with multi-planar reconstruction shows significant LM lesion (arrow). (B) CT angiography of aorta without contrast injection depicts diffuse aortic wall calcification (arrow). (C) Invasive coronary angiography shows significant LM ostial stenosis (arrow).

at the apex and a diastolic murmur also heard in the aortic position with III/VI intensity. Furthermore, a small left cervical lymphadenopathy (short-axis diameter: 10 mm) was detected with no erythema or pain.

Erythrocyte sedimentation rate, C-reactive protein, wright, coombs wright, 2-mercaptoethanol, the venereal disease research laboratory (VDRL) tests, lipid profile, liver function test and thyroid hormone levels were within a normal range. Notably, his hemoglobin level was 12.5 g/dl.

Since some researches have suggested an overlap between tuberculosis and TA, tuberculin test, sputum smear and culture for mycobacterium was carried out, which were all negative.

The echocardiography revealed an ejection fraction (EF) of 30% with global left ventricular (LV) hypokinesia, mild LV enlargement, and moderate mitral, aortic, and tricuspid valves regurgitation. To evaluate the coronary arteries, CT angiography was performed, which determined diffuse media thickening of the aortic root (5 mm), and a significant narrowing of the LM ostium (Fig. 1A). Besides, it displayed evidence of aneurysmal dilation of ascending aorta with extensive aortic wall calcification along its course (Fig. 1B). In this regard, coronary angiography confirmed CT results (Fig. 1C).

CMR examination indicated severe biventricular systolic dysfunction, localized myocardial inflammation and subepicardial fibrosis in the inferior and anteroseptal LV wall in favor of active myocarditis (Fig. 2A). Furthermore, there was evidence of aortic wall thickening (Fig. 2B). In the MRA, aortic arch branches showed mild irregularity and focal narrowing, whereas the celiac trunk (Fig. 2C) and right renal artery had significant stenosis at their origin. In addition late gadolinium enhancement sequence revealed fibrosis of the aortic wall (Fig. 2D).

Considering the mentioned findings (active inflammation of the aortic wall, myocardium, and LM stenosis as well as laboratory findings), all probable differential diagnoses evaluated by an expert rheumatologist, and ultimately the diagnosis of TA considered for the patient.

Regarding the clinical signs, findings of disease activity on CMR in both aorta and myocardial tissue, and decreased EF, intensive medical treatment selected for the patient.

The therapy started with 500 mg of methylprednisolone for 3 consecutive days and then continued at a dose of 0.5 mg/kg, equivalent to 30 mg, daily, and reduced to 10 mg within 6 months.

In addition to steroid therapy, cyclophosphamide started for the patient at a dose of 500 mg for 3 consecutive months. The patient had two children and agreed to take the drug. However,

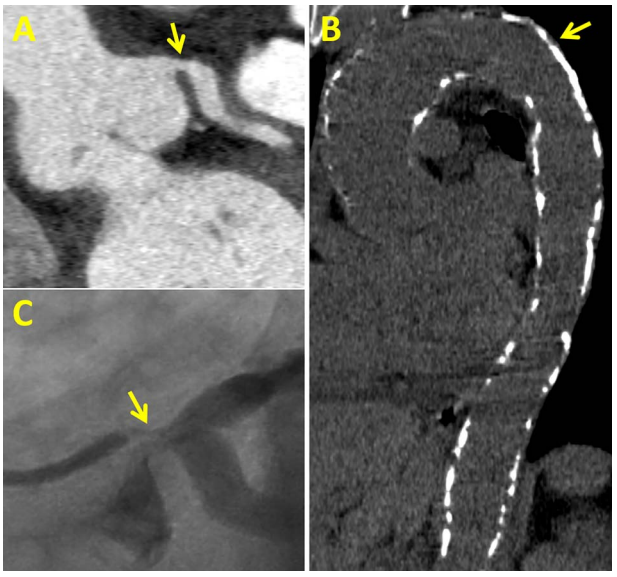

Figure 2: CMR sequences in TA. (A) Late gadolinium enhancement image in candy can view shows gadolinium uptake of the aortic wall. (B) Axial localizer view shows thickening of the aortic wall. (C) MR angiography, sagittal view, depicts aortic wall irregularity (white arrow) and significant stenosis of the celiac trunk ostium (yellow arrow). (D) Late gadolinium enhancement image in basal short-axis view demonstrates LV lateral wall fibrosis.

we sent the patient's sperm to the sperm bank for storage. Subsequently, treatment with injectable methotrexate at a dose of 20 mg per week continued. Due to CHF, anti-TNF drugs were not appropriate for the patient.

DISCUSSION

TA is an idiopathic vasculitis that mainly affects the aorta and its branches, which results in the increased wall thickness of arteries, fibrosis, stenosis, aneurysmal formation or dilation, and thrombosis. In this report, we present an interesting case of TA diagnosed with a multi-modality imaging approach that had a combination of LM coronary artery involvement, entire aortic wall calcification, significant stenosis of the celiac trunk, and myocarditis, which is a rare combination in TA. Signs and symptoms characterizing the vascular inflammatory stage include, but not limited to, fatigue, fever, malaise, palpitation, headache, rash, hemoptysis and weight loss [1]. However, disease activity may persist in the aortic wall, despite the patient being asymptomatic and all inflammatory markers may remain within normal limits. Diffuse aortic wall calcification is a marker of long-standing inflammation in TA, which is a very rare finding [2].

Coronary arteries, like other aortic branches, are involved in TA in 6–30% of cases. The right coronary artery is the most affected one. The most common site of coronary artery involvement is the ostium and proximal segments of the right and left coronary arteries [3]. The optimal coronary revascularization method for Takayasu patients is not known yet. Wong *et al.* concluded that coronary artery bypass grafting (CABG) is better than percutaneous coronary intervention because the restenosis rate and major adverse cardiovascular events are more in PCI procedure than the CABG. More than a 9-fold increased risk of major adverse cardiac events in the percutaneous procedure than CABG reported in this study. On the other hand, CABG in patients with TA has many difficulties [4].

Because the left subclavian and internal mammary arteries are commonly affected, a saphenous vein graft is preferred [3–5].

For the first time, Kuijer *et al*. reported a case in which a Dacron prosthetic patch utilize to insert the proximal anastomosis on an inflamed aorta. The patch prevented contact between inflamed tissue and the graft. They believe that in this way, the risk of graft failure will be reduced [3]. Oishi *et al*. reported successful repair of ostial coronary artery stenosis using a femoral artery patch in Takayasu arteritis. It would be a reasonable surgical method [6].

Therefore, three common surgical approaches are CABG, patch angioplasty of the LM coronary artery ostium, and transaortic coronary ostial endarterectomy. Although surgery is prohibited during the active stage of the inflammation, in a patient with unstable angina, it is applicable.

Hybrid CABG is an alternative technique. Preoperative stent insertion and subsequent off-pump CABG perform in the hybrid method. In the presence of the macroscopic calcification of the ascending aorta, conventional CABG is not the preferred technique [7]. Thus, the Hybrid procedure is useful in patients with porcelain aorta [8, 9]. As an alternative method, percutaneous transluminal coronary angioplasty can perform in Takayasu's disease. This procedure has been offered in high-risk patients who refuse surgery or have severe pulmonary hypertension [8]. Because of the underlying inflammation and the higher rate of in-stent restenosis, interventional therapy was more associated with the re-interventions rate than the surgical treatment [10].

Diffuse aortic wall calcification is a late and rare manifestation of Takayasu arteritis. Multi-modality imaging using cardiac CT and CMR has a pivotal role in the diagnosis of vasculitis and portrays its complications.

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CONFLICTS OF INTEREST STATEMENT

All of the authors mention that they have no conflicts of interest.

AUTHORS' CONTRIBUTION

All authors contributed to the data gathering and writing of the manuscript.

ETHICAL APPROVAL

This study has ethical approval from the ethics committee of Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

GUARANTOR

The Guarantor of this manuscript is Dr Nahid Rezaeian.

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