

Unusual Presentation of Takayasu Arteritis as Acute Myocardial Infarction

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ABSTRACT

Takayasu arteritis is characterized by a granulomatous inflammation that primarily affects the aorta and its branches. The clinical presentation may be varied, depending on vascular conditions. The diagnosis is based on six clinical criteria established by the American College of Rheumatology and confirmed by the presence of three or more of these criteria. The cases of Takayasu arteritis presenting with acute myocardial infarction are rare. This presentation describes the case of a 30 year old patient who was admitted to our hospital with diagnosis of acute anterior wall myocardial infarction, which required primary angioplasty in the anterior descending artery.

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Key words > Takayasu arteritis - Myocardial infarction - Vasculitis

Abbreviations >

ADA	Anterior descending artery	LVSF	Left ventricular systolic function
ECG	Electrocardiogram	AMI	Acute myocardial infarction

INTRODUCTION

Takayasu's arteritis is a granulomatous inflammation that mainly affects the aorta and its large branches. It was described by Dr. Mikito Takayasu (Japanese ophthalmologist) in 1908 as a clinical syndrome characterized by eye complications and accentuated pulse weakness ("pulseless disease"). Although its etiology and pathogenesis are unknown, an autoimmune mechanism is suspected. (1) The cases presenting with acute myocardial infarction (AMI) are rare. This presentation describes the case of a patient who was admitted with an acute coronary syndrome with ST segment elevation and suspected Takayasu Arteritis.

CASE REPORT

A 30-year old woman was derived from another institution with diagnosis of AMI with anterior wall ST-segment elevation and spontaneous reperfusion. She had history of hypertension since her adolescence treated with amlodipine 5 mg/day, and intermittent claudication at 300 meters of 1-month evolution.

On physical examination her left arm blood pressure was 120/60 mm Hg, right arm blood pressure 100/50 mm Hg, and lower limb blood pressure 100/60 mm Hg. Her heart rate (HR) was 75 bpm, respiratory frequency 18 breaths / min and temperature 36.3° C. The patient

had reduced right femoral and humeral pulses, absent in popliteal, posterior tibial and dorsalis pedis regions, and murmur at the right subclavian artery. The rest of the physical examination was normal.

Admission LDH level was 1024 UI/L, with the rest of laboratory tests within normal ranges. ECG at admission showed sinus rhythm HR of 70 bpm, axis 60°, PR 0.20 s, QRS 0.08 s, QT 0.40 s, and negative T in the anterolateral wall

The transthoracic echocardiogram revealed hypokinesia in apical segments and preserved LVSF. Patient outcome was uneventful, and thus, a stress echocardiography was requested showing a significant ischemia in the left anterior descending artery (LAD).

While awaiting for a coronary angiography the patient presented with prolonged class IV angina with anterior subepicardial lesion. Coronary angiography revealed 99% left anterior descending coronary artery (LAD) ostial stenosis (Figure 1A and B). Based on clinical and coronary anatomy an angioplasty with stent in the LAD lesion was performed.

During hospitalization, evaluation was completed with the following additional studies: 1) carotid Doppler ultrasound scan which showed complete left primitive and internal carotid artery occlusion along their full course; 2) angiotomography showing

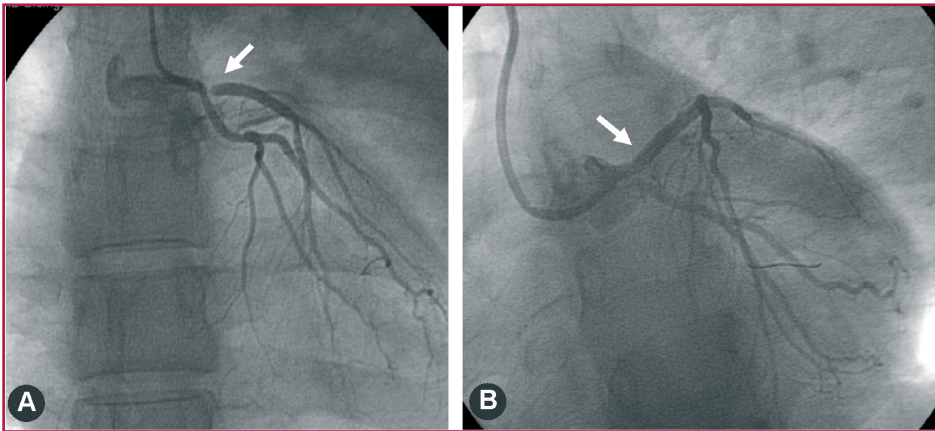


Fig. 1. Coronary angiography presenting critical ostial lesion in the left anterior descending artery with additional thrombus image; preserved distal flow (A). Injection of the left coronary artery post angioplasty with stent (B).

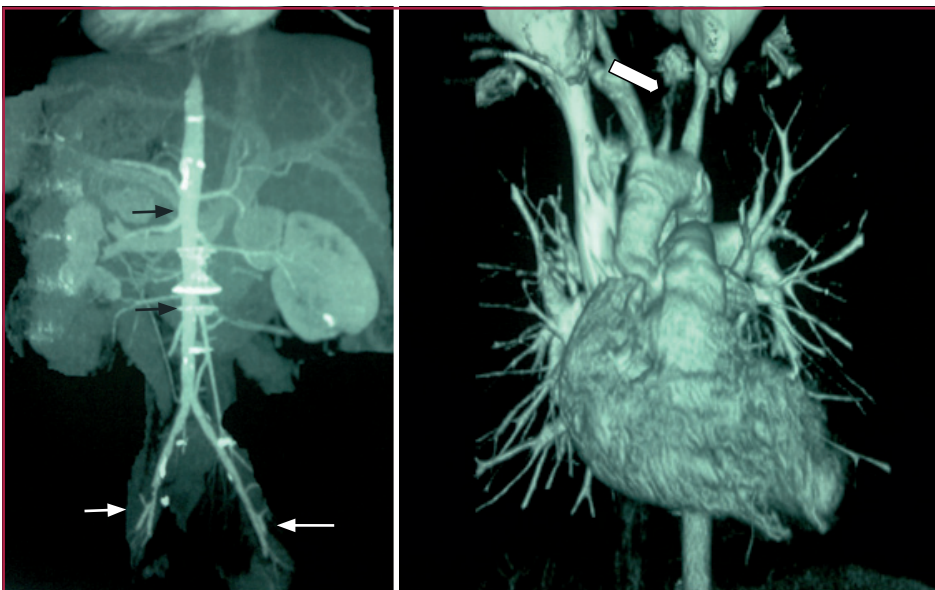


Fig. 2. Angiotomography showing narrow abdominal aorta (black arrows) and severe bilateral obstruction at the level of the iliac arteries (white arrows). The critical injury of the left primitive carotid artery is also seen (white arrow in right image).

total occlusion of the left primitive carotid artery, very narrow abdominal aorta, renal arteries without angiographically significant lesions, and very narrow infrarenal aorta. It also evidenced total occlusion in both external iliac arteries and a severe lesion in the right subclavian artery (Figure 2).

Takayasu arteritis was diagnosed based on the patient's history and vascular involvement detected in the complementary studies and according to the criteria of the American College of Rheumatology.

Within 48 hours she was discharged with aspirin 100 mg/day, methylprednisolone 40 mg/day, clopidogrel 75 mg/day, amlodipine 10 mg every 12 hours, nebivolol 10 mg/day, hydrochlorothiazide 25 mg/day and pantoprazole 40 mg/day.

Currently, the patient is followed up by the Cardiology and Rheumatology outpatient clinics. During outpatient follow-up, evaluation was completed with a resonance angiography of intracranial and extracranial vessels which revealed absence of flow in the left

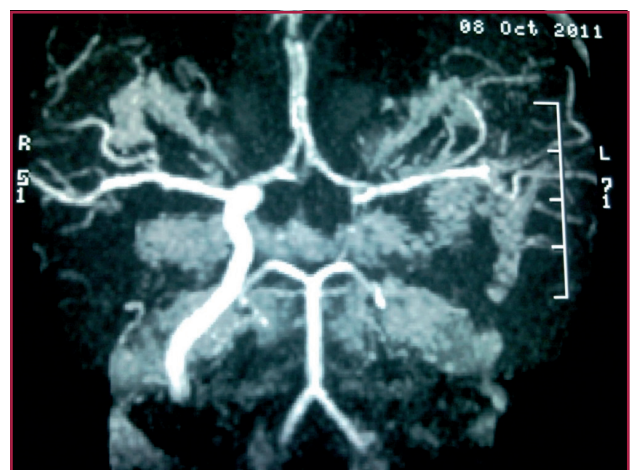


Fig. 3. Magnetic resonance angiography of intracranial and extracranial vessels showing a patent right carotid artery visualized by angiotomography (left), in contrast to the left carotid artery occlusion (right).

intracranial internal carotid, preserved flow in sylvian arteries, which was slightly lower on the left side, preserved anterior cerebral arteries and normal vertebral circuit. The neck vessels showed absence of flow in the left primitive, internal and external carotid arteries. Right carotid arteries and both vertebral arteries were patent (Figure 3).

DISCUSSION

Takayasu's disease is histologically characterized by irregular thickening of the aortic wall and its branches. Early lesions consist of a mononuclear infiltrate of the vascular adventitial vasa vasorum. Later, there may be marked mononuclear infiltrate of the arterial media, accompanied in some cases by granulomatous lesions filled with giant cells and patchy media necrosis. With disease progression even after treatment with steroids, inflammatory reaction is predominantly characterized by collagen fibrosis affecting all layers of the vessel wall (especially the intima) accompanied by lymphocytic infiltration. This is the mechanism by which constrictions of the coronary arteries may lead to AMI. (1)

The American College of Rheumatology established six criteria for the diagnosis of Takayasu arteritis (Table 1). Presence of three or more of these six criteria showed a sensitivity of 92.1% and a specificity of 97%. (1) In our patient 6/6 criteria for the diagnosis of Takayasu's disease were found.

The ratio of women to men was 7.9 to 1 respectively. Most patients (76%) had signs or symptoms attributable to the disease at 40 years or less. However, 24% had no signs or symptoms until after 40 years of age. Hypertension, as in our patient, was the most common sign (69%), followed by diabetes and dyslipidemia. The most common and suggestive symptoms of vascular disease were claudication (39.2%) and dizziness (38.7%).

Complementary studies are variable and depend on the type of clinical presentation (presence of car-

diovascular involvement), ranging from transthoracic and transesophageal echocardiography to peripheral vascular Doppler, coronary angiography, angiography or magnetic resonance angiography. Angiographic findings have shown that aortic disease distribution is: abdominal 63%, descending 57%, ascending 47% and aortic arch 37%. The left common carotid artery (72.1%) is the most frequently affected branch, which is consistent with the findings in our patient, followed by the left subclavian artery (67.1%), right common carotid artery (63.7%) and the right subclavian artery (55.2%). (1, 2)

The treatment, once diagnosed, is based on glucocorticoid monotherapy or associated with immunosuppressants. According to the work of Kerr et al (3) analyzing 60 patients, the use of corticosteroids, alone or in combination with immunosuppressants, showed remission in only 25% of the patients. Of those who had remission, 50% relapsed after treatment.

Shelhaner et al (4) conducted a prospective follow-up of 20 patients for 4.6 years, with glucocorticoid treatment associated or not with cyclosporine. Only 30% responded favorably to treatment based on corticosteroids and 20% required association with cyclosporine. Only two patients of the initial group progressed after 30-48 months.

The Italian group (5) for the study of Takayasu's disease analyzed 104 patients with a median follow up of 15.5 months. Glucocorticoids were the basic treatment; however, 50% of patients required immunosuppressive association. Sixty-nine percent of patients were treated with glucocorticoids or immunosuppressants, 54% of them in an inactive phase of the disease.

Another study followed up 204 patients from 1994-2009, included for presenting at least three of the criteria considered by the American College of Rheumatology for the classification of Takayasu's disease. (6) Patients were divided into two groups: one treated with prednisone and the other treated with prednisone associated with immunosuppressants. However, after

Table 1. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis

Criteria	Definition
Onset at age \leq 40 years	
Claudication of extremities	Development and/or worsening of fatigue and discomfort in muscles of one or more extremities while in use, especially in the upper extremities
Decreased brachial artery pulse	Decreased pulse of one or both brachial arteries
> 10 mm Hg difference in systolic blood pressure between arms	
Bruit over subclavian arteries or aorta	Angiographic narrowing or occlusion of the
Arteriographic evidence of narrowing or occlusion of the entire aorta, its major primary branches or large arteries in the proximal upper or lower extremities	entire aorta, its major primary branches or large arteries in the proximal upper or lower extremities, not caused by atherosclerosis or fibromuscular dysplasia

analysis no significant differences were found.

During the course of the disease, as observed by Kerr et al (3) glucocorticoids were the most used drugs, indicated in 86% of patients, and immunosuppressants were prescribed in 54% of patients.

Upon discharge our patient was treated with glucocorticoids (prednisone 40 mg / day). As disease progressed, the association with cyclosporine was recommended.

CONCLUSIONS

The clinical presentation of patients with Takayasu's arteritis can be as varied as the location of the affected arteries. The cases presented with AMI described in the literature are rare; in the case of our patient it was an anterior AMI.

The patient's prognosis varies depending on which part of the anatomy is affected. The treatments should be focused not only to prevent progression of the disease, but also to improve the quality of life. From the beginning treatment has been based on corticosteroids, which may be associated with immunosuppressants such as cyclosporin.

Early diagnosis and optimal treatment play a key role in the patient's outcome and disease progression.

RESUMEN

Presentación inusual de la arteritis de Takayasu como infarto agudo de miocardio

La arteritis de Takayasu se caracteriza por una inflamación granulomatosa que afecta principalmente la aorta y sus ra-

mas. La presentación clínica puede ser variada, dependiendo de la afección arterial. El diagnóstico se sustenta en seis criterios clínicos, determinados por el American College of Rheumatology, y lo confirman la presencia de tres o más de esos criterios. Los casos de arteritis de Takayasu que debutan con un infarto agudo de miocardio son raros. En esta presentación se describe el caso de una paciente de 30 años que ingresó a nuestro hospital con diagnóstico de infarto agudo de miocardio de cara anterior, que requirió angioplastia primaria en la arteria descendente anterior.

Palabras clave > Arteritis de Takayasu - Infarto del miocardio - Vasculitis

Conflicts of interest:

None declared.

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