

A Case of Severe Advanced Takayasu Arteritis with Acute Myocardial Infarction as First Manifestation

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ABSTRACT: We present the case of a 53-year-old female who presented in the emergency room accusing chest pain, dyspnea to moderate physical strain and physical asthenia. The clinical exam highlighted the absence of pulse in the left upper limb, weak pulse at upper right and lower limbs. The angiography revealed severe coronary lesions, bilateral subclavian occlusion, bilateral renal artery occlusion, infrarenal aortic occlusion. The patient was diagnosed with Takayasu's disease. Sequential interventional revascularization was performed. Takayasu arteritis is a rare disease that can easily be overlooked. It can present with severe coronary lesions which require a complex interventional and medical management.

KEYWORDS: Takayasu arteritis, Myocardial infarction, Percutaneous transluminal coronary angioplasty

Introduction

Takayasu arteritis is a chronic inflammatory, systemic and progressive disease of large arteries. It is a relatively rare disease; it mainly affects the aorta and its major branches, such as brachiocephalic trunk, carotid, subclavian, vertebral, renal, coronary and pulmonary arteries [1,2]. Described for the first time in Japan in 1908, it steadily increased in prevalence in Asia and Oriental countries [3]. Worldwide annual incidence is 1.2-2.6 cases/million inhabitants [2]. It is more frequent among young women, especially in the 2nd or 3rd decade of life (less than 15% of the reported cases developed the disease after the age of 40) [4].

This type of vasculitis is characterized by narrowing, occlusion, or aneurysms of the arteries involved. Clinical presentation is diverse and depends on the affected artery; it could include limb claudication or numbness, vertigo, syncope, visual disturbances, angina pectoris, and convulsions. The distinctive clinical sign is the decrease or absence of the pulse (seen in 80-90% of patients), lower blood pressure in inferior or superior limbs, carotid, subclavian arteries or aortic systolic bruits (in 80-95% of patients), retinopathy, arterial hypertension [2].

The etiology of the disease is still unknown, but the pathological mechanism involves a T cell-mediated autoimmune reaction against arterial wall [5].

We report the case of a patient with Takayasu disease and the difficulties encountered in

addressing this pathology. We have the written consent of the patient for publishing her case.

Case report

We consulted a 53-year-old female that presented to the emergency room complaining of retrosternal pain with posterior irradiation debuted 19-hours before, accompanied by dyspnea to moderate physical strain and asthenia. She was known with the following antecedents: grade 3 arterial hypertension diagnosed at the age of 25, dyslipidemia, obesity, asthma and sero-fibrinous pericarditis diagnosed one month before presentation.

After the clinical exam, the relevant findings were obesity, the impossibility of measuring blood pressure, the absence of the pulse at the upper left and a weak pulse at the upper right and the lower limbs. The ECG revealed sinus tachycardia (116b/min), lack of increase of R wave in V1-4, ST segment elevation of maximum 2mm in V1-4, negative T waves in DI, aVL. Echocardiography showed a mild aortic regurgitation, moderate mitral regurgitation, left ventricular hypertrophy, apical interventricular septum akinesia, antero-lateral wall and anterior wall hypokinesia; LVEF was 35%.

Laboratory analyzes revealed an important inflammatory syndrome (ESR=70mm/h, CRP=33mg/dl, fibrinogen=525mg/dl), neutrophilic leukocytosis, increased myocardial necrosis markers (TnI=3.51ng/ml, CK-MB=51ng/ml), dyslipidemia (LDL=106mg/dl, HDL=38mg/dl).

Based on clinical and paraclinical data, the established diagnosis was: antero-septal ST segment elevation myocardial infarction at

19-hours from onset, Killip I class; arterial hypertension grade 3 with very high cardiovascular risk; moderate mitral regurgitation; mild aortic regurgitation; dyslipidemia; asthma.

Peripheral artery angiography revealed bilateral subclavian artery occlusion, bilateral renal artery occlusion, infrarenal aortic occlusion (Fig.1). Coronary artery angiography revealed chronic left anterior descending artery

occlusion in the first segment, 90% stenosis of the circumflex artery in the first segment, occlusion of an important intermediary branch, and 50% stenosis of right coronary artery (Fig.2A and B).

With the consent of the patient, a dual stage interventional revascularization was performed, first addressing the right subclavian artery, circumflex and intermediate branches of the left coronary artery (Fig.2C). In the second stage angioplasty of the left anterior descending artery was done (Fig.2D).

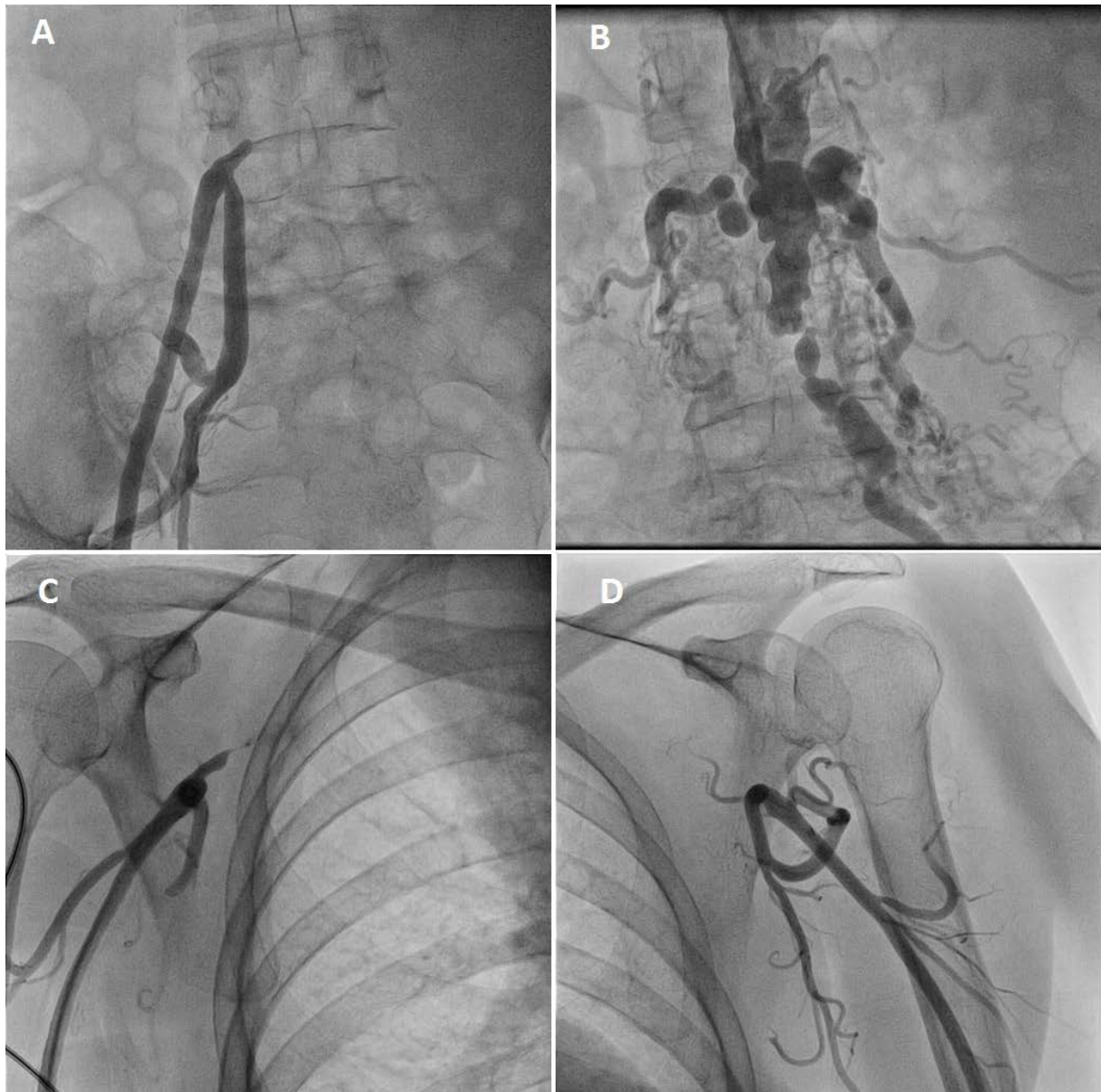


Fig.1. A-D. Arteriography. Occlusion of right iliac artery (A); Occlusion and aneurysms in terminal abdominal aorta and branches (B); Occlusion of right subclavian artery (C); Occlusion of left subclavian artery (D)

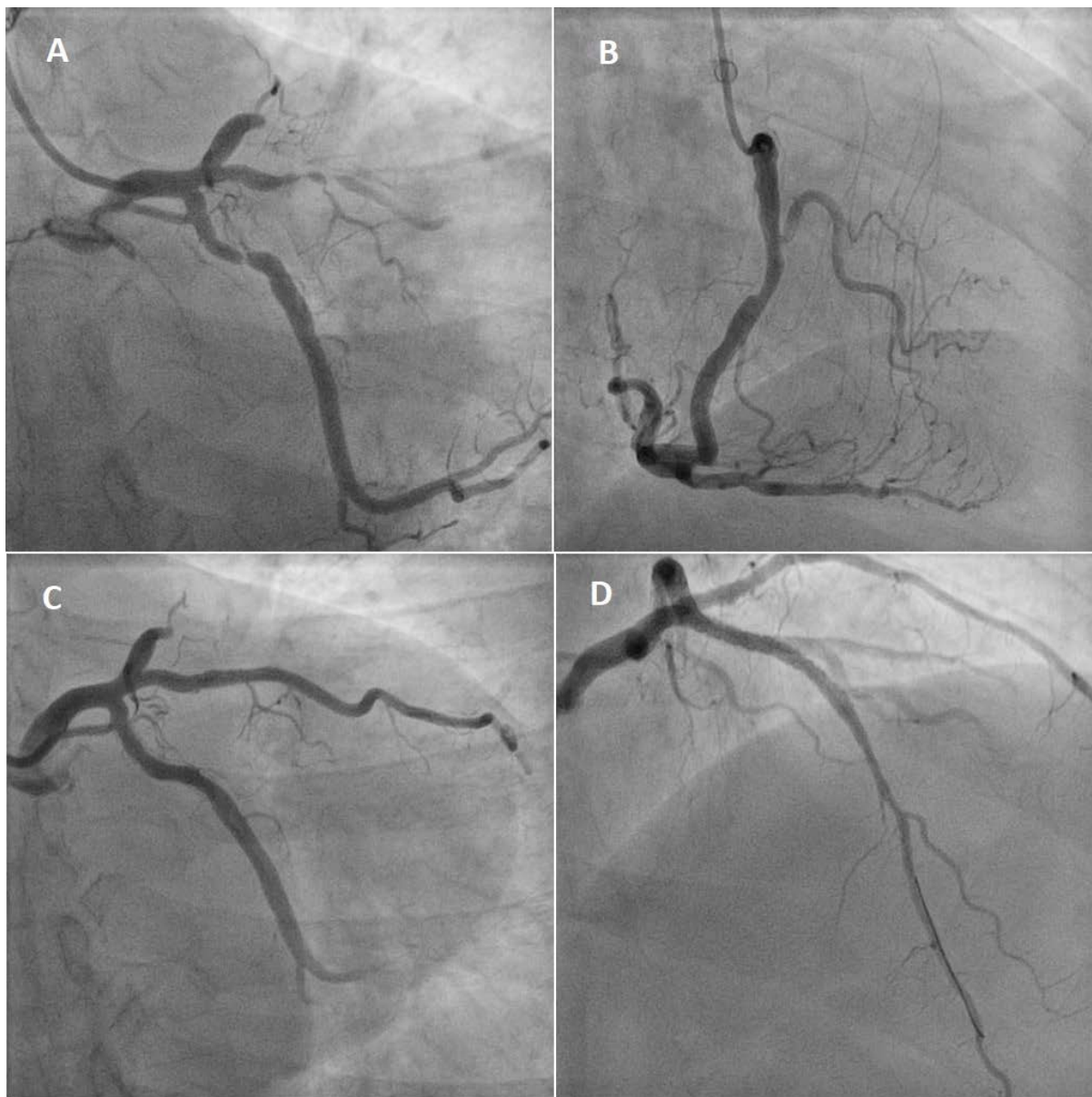


Fig.2. A-D. Coronary angiography. Left coronary artery-occlusion of left anterior descending artery, occlusion of intermediate branch, severe stenosis of circumflex artery (A); Right coronary artery-50% stenosis in mid segment, retrograde perfusion of left coronary (B); Left anterior descending artery after stenting of circumflex and intermediate branches (C); Result after left anterior descending artery stenting (D)

Immediate therapy with beta-blocker, dual anti-platelet therapy, statin, ivabradine, angiotensin conversion enzyme inhibitor was initiated.

Considering clinical and laboratory findings and taking into account the presence of 4 American College of Rheumatology criteria, we diagnosed the patient with Takayasu type V C (+) disease, according to angiographic classification. Female gender is another important element, considering that 80-90% of cases are women. The inflammatory syndrome highlights an active stage of disease.

Following a specialist consultation, the rheumatologist initiated therapy with

methylprednisolone 48mg/day (the dose for active vasculitis). After 2 weeks, the regimen was adjusted by adding methotrexate (20mg/week) and folic acid, and since the disease was considered severe due to the biological activity (non-specific inflammatory syndrome, high ESR, and increased CRP despite Corticosteroid therapy), and due to the presence of complications (obstruction of the subclavian arteries), the dose of methylprednisolone was increased to 64mg/day.

According to recent studies, the most effective therapy is immunosuppressive therapy, namely anti-TNF-alpha and anti-IL-6. Because these are currently unavailable in our country,

Methotrexate was preferred, with other advantages being the single-dose use, and moderate long-term toxicity. This was initiated quickly because the effect only occurs after 8-12 weeks.

Discussion

The diagnosis of Takayasu disease in our patient was established using the current criteria

of the American College of Rheumatology (Table 1). Because we are dealing with a relatively young woman with many associated cardiovascular risk factors, it is a question of elucidating the etiology of the multiple disorders seen in this patient. Unfortunately, the case was diagnosed late and accidentally.

Table 1. The American College of Rheumatology criteria for the classification of Takayasu arteritis [6]

Criteria	Description
Age at disease onset <40 years	Development of symptoms or findings related to Takayasu arteritis at age <40 years
Claudication of extremities	Development and worsening of fatigue and discomfort in muscles of one or more extremity while in use, especially the upper extremities
Decreased brachial artery pulse	Decreased pulsation of one or both brachial arteries
BP difference >10mmHg	Difference of >10mmHg in systolic blood pressure between arms
Bruit over subclavian arteries or aorta	Bruit audible on auscultation over one 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 said to have Takayasu arteritis if at least 3 of these 6 criteria are present. The presence of any 3 or more criteria yields a sensitivity of 90.5% and a specificity of 97.8%	

Given the fact that the patient was known with early onset hypertension, a more thorough evaluation could have been decisive in a prompt diagnosis of Takayasu disease. It was probably considered that hypertension is most likely essential, given the family history of hypertension (father). Measuring blood pressure at both arms is very important, considering that its absence in the left arm has not been observed in our patient. A decreased elasticity of the aorta due to the inflammatory process and fibrosis may be the cause of secondary hypertension in

this case. Due to the chronic ischemia, a rich collateral circulation has developed as adaptive mechanism. Thus, the collateral vessels limiting ischemia played an important role in maintaining the viability of affected tissues, and this could explain the mild symptoms expressed by the patient prior to the hospitalization.

Another important point in our patient is the extensive coronary involvement. In the angiographic classification of Takayasu disease, coronary artery involvement is designated as C (+) (Table 2).

Table 2. Angiographic classification of Takayasu arteritis [7]

Type	Vessel involvement
Type I	Branches from the aortic arch
Type II a	Ascending aorta, aortic arch and its branches
Type II b	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 II b and IV
Involvement of the coronary or pulmonary arteries should be designated as C (+) or P (+), respectively	

The incidence of coronary artery disease has been reported to be around 10%, with 3 types of lesions: type 1, stenosis of the coronary ostia or proximal segments of the coronary arteries; type 2, diffuse or focal coronary arteritis; and type 3, coronary aneurysm [7]. Type 1 is most frequent. The recommended treatment of Takayasu's arteritis with coronary stenosis is percutaneous angioplasty or surgery, but there is a high risk of restenosis.

Considering the presence of numerous complications, we wanted to monitor the patient every 3 months. In terms of prognosis, this case had an unfavorable one given the many cardiovascular complications. This was also underlined by Park et al [4], who demonstrated that survival rates at 10 years are about 36%.

Of major importance in the management of Takayasu patients is the assessment of disease activity, through clinical and paraclinical examinations: biological (CRP, ESR) and imaging (CT, MRI, coronary angiography) [8]. Recent studies show that Pentraxin-3 dosing has increased specificity in assessing disease activity, compared to general inflammation markers. Among the newer imaging techniques, positron emission tomography is useful for detecting Takayasu arteritis in the subclinical stage.

Conclusion

Takayasu arteritis is a rare disease that can easily be overlooked, especially if you do not insist on a thorough clinical examination and on the occurrence of secondary causes of hypertension in young patients. Early diagnosis and treatment, associated with aggressive correction of risk factors and adherence to treatment of the patient can improve the prognosis of the disease. Acute myocardial infarction can be the initial manifestation. Coronary artery involvement can be severe, with multivessel disease, requiring complex techniques of interventional revascularization.

Abbreviations

ESR-erythrocyte sedimentation rate;
CRP-C-reactive protein;
TnI-troponin I;
CK-MB-creatinine kinase MB;
LVEF-left ventricular ejection fraction;
CT-computed tomography;
MRI-magnetic resonance imaging;

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