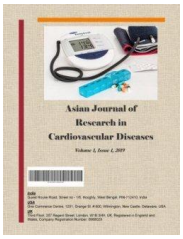

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Cardiac Manifestations of Takayasu Disease: A Case Report and a Review of the Literature

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Takayasu's disease is a rare chronic inflammatory vasculitis, preferentially affecting young women with predominance on the aorta and its first branches. We report the case of a left ostial common trunk involvement during a hypertensive emergency such as ST-negative acute coronary syndrome associated with aortic insufficiency revealing the disease and we will detail its various cardiovascular manifestations.

Keywords: Takayasu disease; hypertensive emergency; ST-negative acute coronary syndrome; aortic insufficiency.

1. INTRODUCTION

Takayasu's disease is a chronic inflammatory arteritis of unknown etiology, preferentially affecting young women, called "pulseless women's disease", with segmental involvement of the aorta and its main branches. Thickening of the vascular wall is the most characteristic early sign of the disease, progressively leading to stenosis, thrombosis and sometimes to the development of aneurysms. We report the case of a left coronary ostial common trunk involvement revealing Takayasu disease.

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2. CASE PRESENTATION

A female aged, young woman of 40 years old, without particular cardiovascular risk factors. Admitted for intense dizziness and angina chest pain, constrictive retro sternal, radiating to the left upper limb. Clinical examination on admission noted normally colored conjunctiva, tachycardia at 100 bpm with BP at 200/105 mmHg Cardiac examination found regular heart sounds without peripheral signs of heart failure. The pleuropulmonary examination was unremarkable as was the rest of the somatic examination. The basal resting electrocardiogram showed an SRR of 100 beats/min an extended anterior and inferior ST-segment shift, and an AVr ST-segment shift (Fig. 1).

The biological workup showed troponins I at 90 times normal. Resting transthoracic echocardiography shows a dilated LV, asymmetric global hypokinesis with impaired left ventricular systolic function (EF estimated at 45% by Simpson biplane). Left ventricular filling pressures are not elevated with Moderate Aortic Insufficiency. In addition, there was no pericardial effusion or intra-cavity thrombus. A brain CT performed without abnormality as well as an FO performed does not show any abnormality. The diagnosis of high risk NON STEMI ACS was retained and a pharmacological treatment based on Clopidogrel, Enoxaparin, Aspegic, Morphine, and Nicardipine injection was initiated in the emergency room, then referred to the interventional cardiology unit for coronary angiography. The coronary angiography showed an isolated subocclusive stenosis of the ostium of the common trunk of the left coronary artery (Fig. 2).

The right network was free of lesions (Fig. 3). The patient underwent emergency coronary artery bypass surgery with aortic valve replacement with simple postoperative follow-up. Pathological examination of the internal mammary artery biopsy showed lesions suggestive of Takayasu disease. The echodoppler of the supra-aortic trunks showed a parietal thickening of the right internal carotid artery and the subclavian artery, which was homogeneous and circumferential, confirming Takayasu disease. The patient was discharged on the following medical treatment: Clopidogrel 75 mg/d, Aspirin 75 mg/d, Beta-blocker 2.5 mg/d, ACE inhibitor 5 mg/d and Rosuvastatin 20 mg/d, Prednisone 0.5mg/kg then referred to the Internal Medicine department for follow-up.

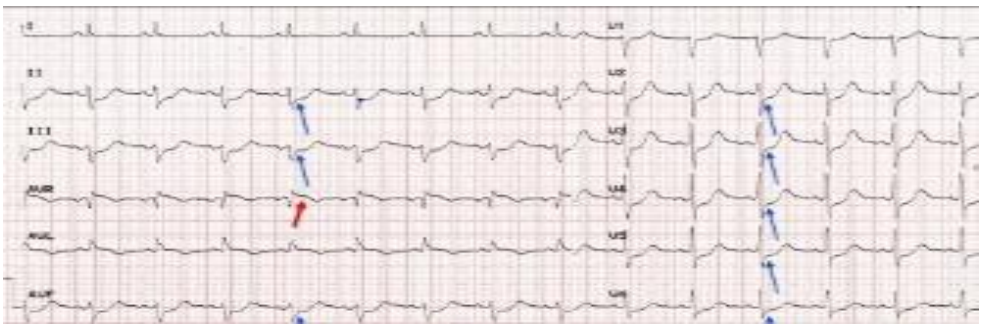

Fig. 1. Extended anterior ST undershift, inferior with Avr overshift

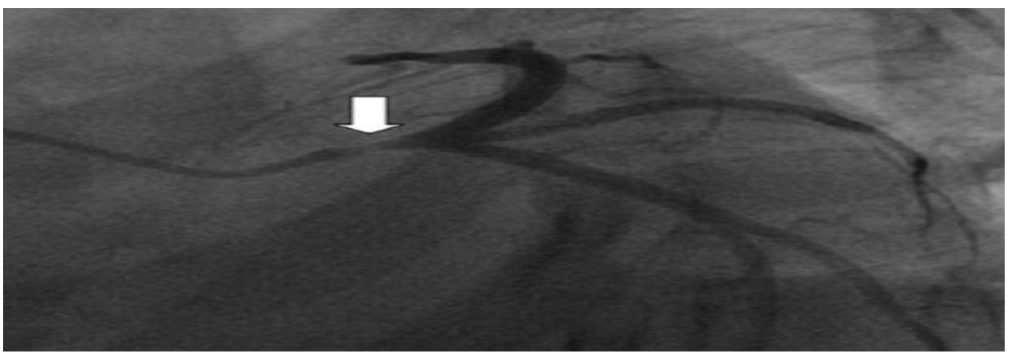

Fig. 2. Caudal OAG incision showing subocclusive stenosis of the ostium of the common trunk of the left coronary artery (arrow)

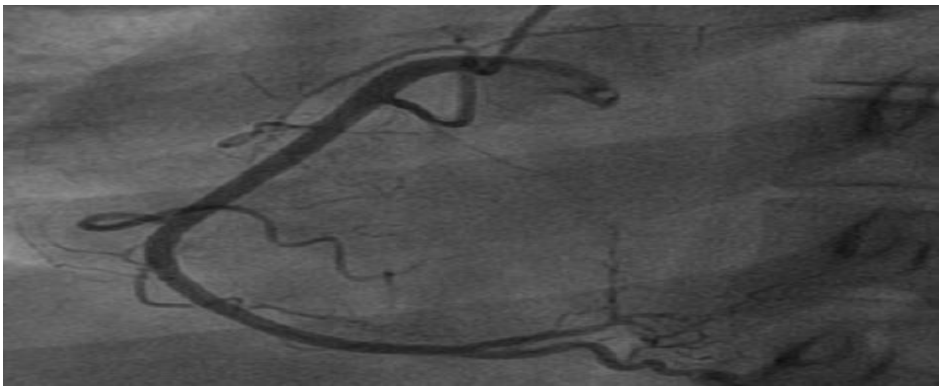

Fig. 3. Incidence OAG showing a right coronary network free of lesions

3. DISCUSSION

Takayasu's disease is a chronic inflammatory arteritis of unknown origin that affects large vessels, mainly the aorta and its main branches. The prevalence of the disease is higher in Japan (40 per million populations), Latin America and Africa with an annual incidence of 2 to 3 cases per million populations. It is a disease of young subjects occurring during the 2 or 3 decades with a female predominance (62% to 97% of patients depending on the studies). The cause of the disease remains unknown, but a few cases of familial involvement have been described. One study [1] identified the genes coding for interleukins 12B, IL 2 and IL 6 as susceptibility loci. In most series, there is a high prevalence of proven tuberculosis [2], and the Th1 and Th17 immune response seems to play an important role in the activity of the disease [3]. Clinically, it is classic to distinguish the acute period called pre-occlusive from the occlusive phase characterized by ischemic manifestations. The pre-occlusive or systemic phase associates general signs, skin signs (erythema nodosum), pain on the arterial pathways and sometimes ophthalmological involvement: episcleritis, anterior uveitis. The occlusive period or vascular phase is the consequence of arterial lesions (stenosis, obliteration, aneurysm). Cardiac involvement is found in 30 to 40% of cases and is considered one of the criteria for the severity of the disease [4]. The frequency of coronary involvement varies according to the series (5 to 45%). It results from ostial involvement as found in our clinical presentation or proximal involvement and is most often manifested by angina; this involvement is often associated with aortitis which may be responsible for an aortic leak as shown in our case, where the aortic leak may be the etiology and an involvement of the subclavian arteries and sometimes poses a therapeutic problem in terms of revascularization [5]. Clinical myocardial involvement is rare but perfusion abnormalities are frequently observed on thallium scintigraphy (84%) and late gadolinium enhancement on MRI (26%) without coronary involvement [6]. Aortic leakage has been reported in 13 to 25% of cases [7], which is confirmed in our patient who presented a probably chronic middle aortic leakage due to the dilatation of the left ventricle. This aortic leakage may be secondary to annular dilatation following an aneurysm of the ascending aorta, or by retraction of the aortic cusps, but the combination of the two mechanisms remains the most frequent in the context of aortitis. Aortic valve replacement (AVR) is the only therapeutic alternative to correct aortic leakage during this disease, which is done in our patient based on the recommendations of learned societies. It will improve the left ventricular function [8], however, some complications may be observed postoperatively and require revision surgery, in particular prosthetic disinsertions, pseudo aneurysms and endocarditis on prosthesis due to tissue fragility and inflammation. Arterial hypertension is very frequent in Takayasu's disease and can be due to various etiologies: renal arterial involvement, aortic pseudo coarctation and parietal stiffness secondary to vascular involvement, widening of the differential in case of aortic insufficiency which is found in our patient who presented for a hypertensive emergency revealing a coronary syndrome.Intermittent claudication of the lower limbs may reveal the coexistence of stenosis and dilatation or aneurysm of the thoracic or abdominal aorta, which are very suggestive of the disease, especially when the vascular wall is thickened. Involvement of the digestive vessels (celiac trunk and mesenteric arteries) is quite frequent, but the occurrence of mesenteric angina is rare. Finally, renal artery stenosis is

frequent and responsible for renovascular hypertension. The diagnosis is based on imaging. Currently, echo-Doppler, CT angiography and nuclear magnetic resonance imaging are reliable and rapid methods of assessing the lumen and the vessel wall. Quality of life is severely impaired in Takayasu disease, while the prognosis is generally good. Diagnosing TA relies on clinical presentation, characteristic structural arterial abnormalities, and evidence of inflammatory vasculopathy on imaging or histology [9]. The degree of arterial lesions and the extension of vascular involvement usually determine the severity of clinical manifestations [10]. The main causes of death are heart failure, stroke, renal failure and persistent inflammatory syndrome. Percutaneous transluminal angioplasty and sometimes revascularization surgery are necessary in case of critical ischemia. Corticosteroid therapy is the first line treatment, in case of failure the addition of methothrexate would allow to control the disease [11; 12].

4. CONCLUSION

The takayastu disease is a rare pathology which can enter in the framework of the autoimmune diseases often under diagnosed the cardiac attacks frequency of the most frequent cardiac attacks are the aortic valvulopathy and the coronary disease and their occurrence conditions in part the prognosis of the disease.

DISCLAIMER

The products used for this research are commonly and predominantly use products in our area of research and country. There is absolutely no conflict of interest between the authors and producers of the products because we do not intend to use these products as an avenue for any litigation but for the advancement of knowledge. Also, the research was not funded by the producing company rather it was funded by personal efforts of the authors.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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