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Case report

Takayasu Arteritis: A Case Report

Arteritis de Takayasu: a propósito de un caso

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ABSTRACT

Takayasu arteritis has a multifactorial etiopathogenesis. Both genetic and infectious factors are implicated in its development. The diagnosis of this disease is often delayed due to its nonspecific and general symptoms, which can lead to misinterpretations. It can affect the coronary arteries. Coronary angiography via axial computed tomography (CT) is the diagnostic technique of choice. In many cases, it is discovered in patients who present with symptoms such as chest pain and effort-induced dyspnea. A case is presented of a 46-year-old Caucasian woman in whom coronary computed tomography angiography revealed the presence of aneurysms in the left main coronary trunk, the left anterior descending artery, and the circumflex artery in its proximal segments, along with multiple arteriovenous fistulas along their courses. The findings were interpreted as Takayasu arteritis.

Keywords: Takayasu arteritis, coronary angiography, axial computed tomography

RESUMEN

La arteritis de Takayasu posee una etiopatogenia multifactorial. En la misma se invocan factores genéticos e infecciosos. Frecuentemente se realizan múltiples interpretaciones antes de llegar al verdadero diagnóstico de esta enfermedad porque presenta síntomas generales e inespecíficos que pueden llevar a la confusión. Puede afectar las arterias coronarias. La angiografía coronaria por tomografía axial computarizada es la técnica de elección para diagnosticarla. En muchas ocasiones resulta un hallazgo en pacientes que refieren síntomas como dolor torácico y disnea de esfuerzo. Se presenta un caso de una mujer de 46 años de edad, de raza blanca, que en la angiografía coronaria por tomografía axial computarizada se encontró la presencia de aneurismas en el tronco coronario izquierdo, la arteria descendente anterior y la arteria circunfleja en sus segmentos proximales y con múltiples fístulas arteriovenosas en sus trayectos que se interpretó como arteritis de Takayasu.

Palabras clave: Arteritis de Takayasu, Angiografía coronaria, Tografía axial computarizada

INTRODUCTION

Takayasu arteritis is a multifactorial disease with an etiology that remains incompletely understood. It has been recognized since 1908, when Japanese ophthalmologist Mikito Takayasu reported vascular ocular alterations in a 21-year-old female patient, characterized by peculiar arteriovenous anastomosis at the optic disc, caused by retinal ischemia secondary to large vessel vasculitis.[1] Later, in 1948, Shimizu K and Sano K[2] described the clinical features of the disease, which currently do not include ophthalmic findings.[2] The first case outside of Japan was reported in 1952 in Ohio.[3]

Although the etiology of the disease remains incompletely understood, evidence from multiple studies over the last decade indicates a close association with genetic and infectious factors. Additionally, autoimmune alterations mediate its etiology, with a clearly understood role of cellular immunity, while the role of humoral immunity remains less defined.[4-8]

The geographical distribution and prevalence of Takayasu arteritis vary across regions. In countries such as India, Korea, Japan, and other Southeast Asian nations, it is more frequently identified as a cause of renovascular hypertension.[8,9] In contrast, in North America, fibromuscular dysplasia and atherosclerosis are more common causes of renovascular hypertension,[10,11] confirming the genetic link. Furthermore, its predominance in females has led to the proposal of a hormonal theory in its etiopathogenesis.[12]

Takayasu arteritis has also been reported in regions of South America, Africa, Europe, and North America, although its genesis is theoretically attributed to Asia.[13-15] These differences may be due to the fact that the disease is not commonly considered in the diagnostic thinking of healthcare providers or due to the lack of access to coronary computed tomography angiography for diagnosis.

The natural history of Takayasu arteritis is characterized by variability, as there are no pathognomonic signs Thus, the onset of inflammatory or systemic symptoms may precede the presentation of vascular symptoms, which can delay the diagnosis.[16,17] Frequently, multiple interpretations are made before reaching the correct diagnosis due to the presence of general and nonspecific symptoms that can cause confusion.[8,14]

The acute phase is clinically characterized by symptoms such as night sweats, anorexia, and weight loss, whereas the chronic phase, systemic manifestations appear depending on the affected target organs, including claudication of the upper and lower limbs, pulse asymmetry, arterial hypertension, and ischemic symptoms.[18] Timely diagnosis significantly improves the prognosis due to the assurance of appropriate and prompt management.[8,19]

Takayasu arteritis is most frequently diagnosed at approximately 39 years of age in White patients and around 30 years in Black patients.[20] This vasculitis is characterized by inflammation of all arterial layers (intima, media, and adventitia) of large vessels, particularly those in the neck, thorax, and abdomen, although it can also involve smaller arteries, such as the coronary arteries.

The development of lesions in the coronary arteries, interpreted as Takayasu arteritis, typically occurs in the proximal segments of the coronary arteries, with a higher incidence in the left anterior descending artery, followed by the right coronary artery.

Coronary computed tomography angiography is the diagnostic technique of choice and often detects these lesions in patients presenting with symptoms such as chest pain and exertional dyspnea. Another diagnostic test is coronary angiography, but it is more invasive. Meanwhile, multi-detector computed tomography scanners has further enhanced diagnostic capabilities due to its superior technical quality.[21]

CASE REPORT

A case is presented of a 46-year-old Caucasian woman, with a history of hypertension, who reports exertional dyspnea and oppressive retrosternal chest pain radiating to the neck and back. This pain is also accompanied by a rapid decline in her functional capacity, particularly with physical exertion. The patient also complains of palpitations.

During the exercise stress test, electrical changes were observed, characterized by ST-segment depression in the precordial leads, with a delayed recovery following exercise.

Laboratory tests included markers for myocardial necrosis, which were found to be within normal limits.

The echocardiogram performed revealed the following findings: discreet remodeling of the left ventricle with segmental contractility dysfunction, characterized by hypokinesia of the anterior septal, apical septal, anteroapical, and apical segments, with mild impairment of apical closure; left ventricular diameters at the upper limit, approaching dilation; mild concentric left ventricular hypertrophy; the aortic valve showed no alterations, while the mitral valve displayed mild thickening of the posterior leaflet, with prolapse of the anterior leaflet and very mild functional central regurgitation; the right heart chambers and their valvular apparatus were normal; and no evidence of pulmo-

nary hypertension or other abnormalities was detected.

Coronary computer tomography angiography and calcium score were performed. The latter showed a score of zero. Coronary computer tomography angiography revealed the presence of aneurysmal dilatations in the left main coronary trunk, left anterior descending artery, and circumflex artery in its proximal segments (**Figure 1**). In the case of the left anterior descending artery, the dilatations extended to the mid-segment and included multiple arteriovenous fistulas towards the anterior interventricular vein (**Figure 2**). The circumflex artery was affected in its proximal segment and also showed arteriovenous fistulas toward the great cardiac vein (**Figure 3**).

Functional coronary analysis showed that the myocardial tissue was well perfused in both phases of the cardiac cycle, with preserved ventricular function, as the aneurysmal dilatation did not affect blood flow. No intracoronary plaques were present. The arteriovenous fistulas led to dilatation of the coronary venous system, combined with variable degrees of stenosis.

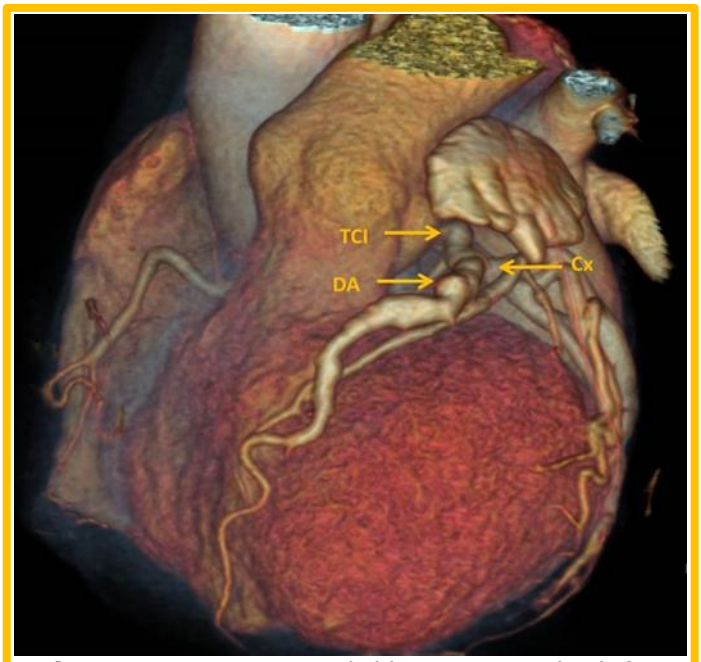

Figure 1 – Aneurysmal dilatations in the left main coronary trunk (TCI), the left anterior descending artery (DA) and the circumflex artery (Cx) in its proximal segments**.**

COMMENT

Cardiac computed tomography is a first-line technique for diagnosing anatomical alterations of the heart and coronary vessels, and has therefore become the benchmark test for studying these conditions. The presence of occlusive and non-occlusive coronary abnormalities is easily visualized, particularly aneurysmal dilatations with the presence of fistulas, as in the presented case, diagnosed as Takayasu arteritis with its typical angiographic alterations.[22]

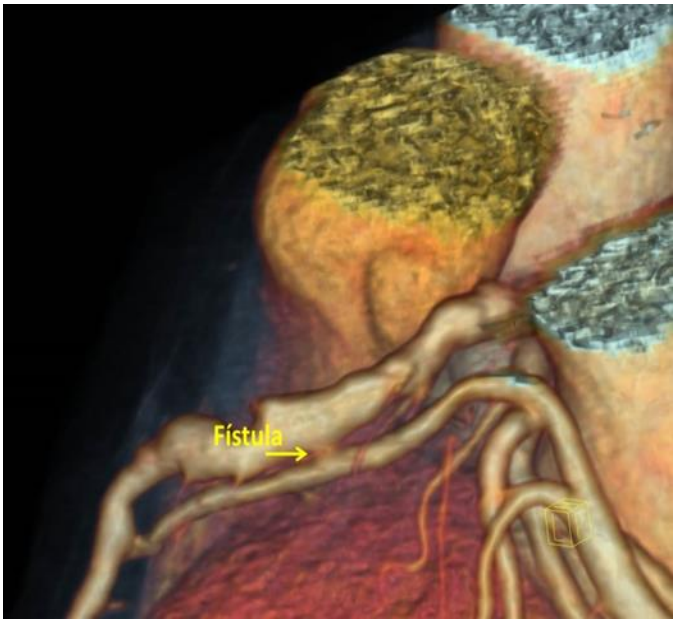

Figure 2 – Aneurysmal dilatation extending to the mid-segment of the left anterior descending artery and multiple arteriovenous fistulas toward the anterior interventricular vein.

Although diagnostic confirmation of Takayasu arteritis is based on these images, as a vasculitis, there are inflammatory markers that may be elevated, such as C-reactive protein and erythrocyte sedimentation rate.[23]

In a series of autopsies, coronary aneurysms were found in 1.4%[24] of cases, while conventional coronary angiography reports an incidence of 4.9%[25], and coronary CT angiography shows 2.7%.[26]

Coronary aneurysms are those segments of a vessel with a diameter 1.5 times greater than the unaffected segment or the coronary vessel with the largest diameter. By definition, they must affect less than 50% of the total length of the artery, distinguishing them from coronary ectasia.[27]

They are classified based on the vascular wall into true aneurysms and false aneurysms, and according to their morphology, into saccular or fusiform.[26,28,29] Additionally, according to their size, they are referred to as giant aneurysms if they measure more than 8 mm in children or more than 20 mm in adults.[30,31]

Generally, coronary aneurysms are clinically silent and are often diagnosed incidentally. When symptomatic, they present with signs similar to those of obstructive coronary artery disease. The prognosis of aneurysms is generally favorable when the underlying cause is properly managed, although it is suggested that

their presence may be a predictive factor for mortality.[32]

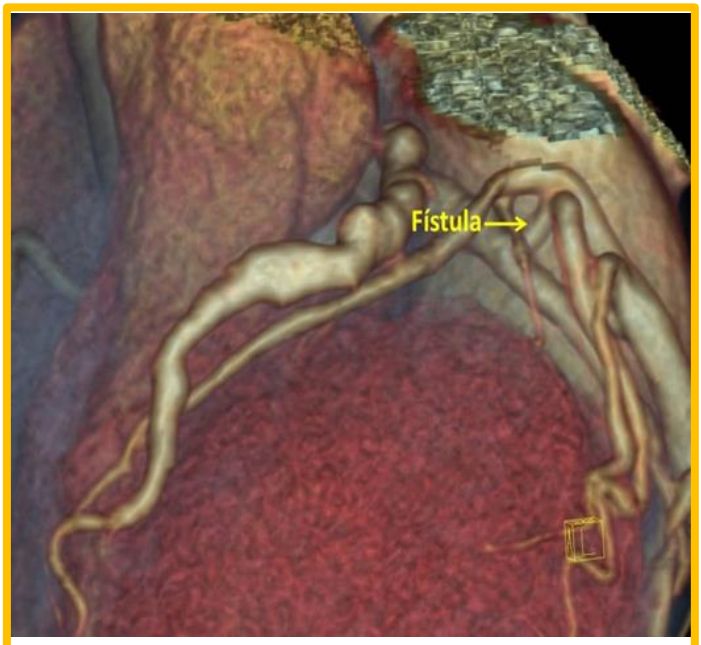

Figure 3 – Aneurysmal dilatation of the circumflex artery in its proximal segment and arteriovenous fistulas toward the great cardiac vein.

On the other hand, coronary fistulas can be either congenital or acquired anomalies, with the latter more frequently reported in patients with Takayasu arteritis and severe coronary atherosclerotic disease. The most frequently described fistulous pathways are between coronary arteries themselves, between arteries and coronary veins, and between coronary arteries and cardiac chambers or the pulmonary artery trunk.[33,34]

The prognosis of Takayasu arteritis is favorable in the short term, but its progressive evolution can lead to vascular complications that increase mortality up to 27% after ten years.[35]

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