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Trombosis aórtica en una mujer con arteritis de Takayasu, ¿manifestación inusual o compromiso paraneoplásico? Reporte de caso

Aortic thrombosis in a woman with Takayasu arteritis, unusual manifestation or paraneoplastic involvement? Case report

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Palabras clave: arteritis de Takayasu; vasculitis sistémica; trombosis; neoplasias mamarias; anticoagulantes; inmunosupresores.

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RESUMEN

La arteritis de Takayasu es una vasculitis de grandes vasos que afecta la aorta y sus principales ramas, caracterizada por síntomas sistémicos inespecíficos y anomalías vasculares, principalmente estenosis o enfermedad aneurismática. La arteritis de Takayasu carece de una asociación clara con eventos trombóticos. El vínculo poco frecuente entre cáncer y vasculitis destaca la necesidad de una evaluación exhaustiva de los trastornos procoagulantes en pacientes con arteritis de Takayasu y trombosis. Presentamos un caso único de trombosis como manifestación inicial de arteritis de Takayasu.

ABSTRACT

Takayasu arteritis is a type of large vessel vasculitis that affects the aorta and its major branches and is characterized by nonspecific systemic symptoms and vascular abnormalities, primarily stenosis or aneurysmal disease. Takayasu arteritis lacks a clear association with thrombotic events. The rare link between cancer and vasculitis highlights the need for a thorough evaluation of procoagulant disorders in Takayasu arteritis patients with thrombosis. We present a unique case of thrombosis as the initial manifestation of Takayasu arteritis.

INTRODUCTION

Takayasu arteritis (TA), first described by Mikito Takayasu, in 1908 and known as "pulseless disease," is a type of large-vessel vasculitis characterized by chronic granulomatous inflammation. This inflammation primarily involves major arteries, causing stenosis or aneurysmal disease[1]. TA predominantly affects the aorta and its primary branches, but isolated cases involving the pulmonary artery or coronary circulation have also been reported[2]. Epidemiological studies have demonstrated significant variability in prevalence, ranging from 0.9-360 cases per million inhabitants[3].

CLINICAL CASE

A 48-year-old woman with a history of active smoking presented to a healthcare center with nonspecific abdominal pain persisting for one month. Thoracoabdominal angiotomography

revealed two thrombi: one in the thoracic aorta (Figure 1) and another in the superior mesenteric artery (SMA) at its origin (Figure 2). Anticoagulation with unfractionated heparin was initiated, and the patient was referred to a higher-level medical center for further evaluation.

Upon admission to our institution, the patient was in good general condition with stable vital signs and no clinical signs of new thromboembolic events or ischemic progression. A systemic review revealed significant unintentional weight loss over the preceding three months, claudication of the upper limbs, hyporexia, and asthenia.

A vascular examination revealed a systolic murmur over the abdominal aorta and left supraclavicular region, along with decreased intensity of the left brachial and radial pulses. Laboratory analysis revealed moderate microcytic anemia and leukocytosis with neutrophilia, with no other abnormalities detected (Table). Cardiac embolism and antiphospholipid syndrome were ruled out, leading to a suspicion of TA.

A review of the external thoracoabdominal CT angiography confirmed vascular involvement of the aorta and SMA without additional abnormalities. To further evaluate arterial compromise, including the supra-aortic vessels, an angioresonance scan was performed (Figure 3). This study revealed eccentric stenosis of the left subclavian artery, thickening of the left vertebral artery, and active inflammation in the SMA. Based on these findings, the initial differential diagnoses considered included fibromuscular dysplasia, atherosclerosis of medium- and large-sized vessels, segmental arterial mediolysis, and other non-inflammatory vasculopathies. However, the lesion distribution, clinical context, and presence of active inflammation supported a vasculitic process as the most likely diagnosis. These findings fulfilled the ACR/EULAR classification criteria for TA (15 points). Consequently, treatment was initiated with intravenous methylprednisolone pulses (500 mg/day for three days), followed by oral prednisone (50 mg/day).

The presence of arterial thrombosis was striking and uncommon in patients with large-vessel vasculitis, prompting an evaluation to rule out malignancy. During screening, a breast ultrasound revealed a 20×13×10 mm nodular lesion (BI-RADS 4C), suggestive of infiltrating breast carcinoma. A rereading of the angioresonance confirmed a solitary nodule in the right breast. Staging chest and abdominal tomography revealed no evidence of metastatic disease (Figure 4). Pathology and immunohistochemistry studies were performed, and outpatient management was initiated with prednisone 50 mg daily (gradually tapered), azathioprine 50 mg and enoxaparin 60 mg every 12 hours. Immunohistopathological analysis confirmed an infiltrating ductal carcinoma of the breast that was positive for CK7, GATA3, estrogen receptor, and progesterone receptor but negative for HER2. Neoadjuvant chemotherapy with doxorubicin and cyclophosphamide, followed by paclitaxel, was administered, and a favorable oncological response was achieved.

Figure 1: Contrast-enhanced computed tomography of the chest and abdomen. The green arrow indicates a 2.8 mm thick thrombus adhered to the left aortic wall. The blue arrow indicates a pedunculated thrombus measuring 5 cm in length with an extension of 14 mm into the right vascular lumen.

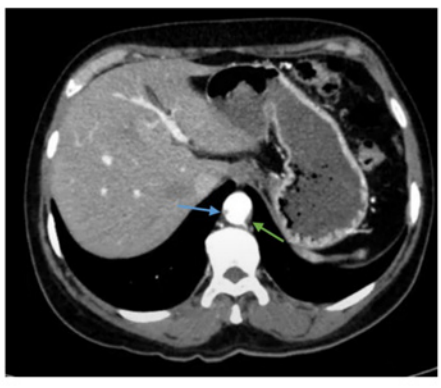

Figure 2: Abdominal computed tomography, reconstruction in arterial phase. **A)** and **B):** the arrow indicates occlusion of the superior mesenteric artery by a thrombus in the proximal and middle third with poor distal re-canalization.

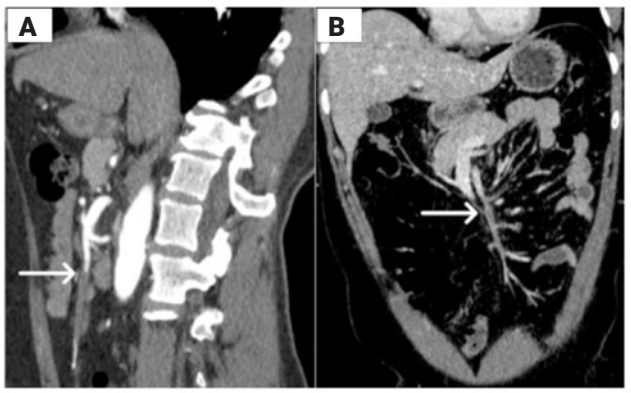

Figure 3: Angiography of the aortic and supra-aortic vessels. **A)** The arrow indicates stenosis at the level of the left common carotid artery. **B)** The arrow indicates stenosis of the left subclavian artery 7 mm from its origin with adjacent wall thickening.

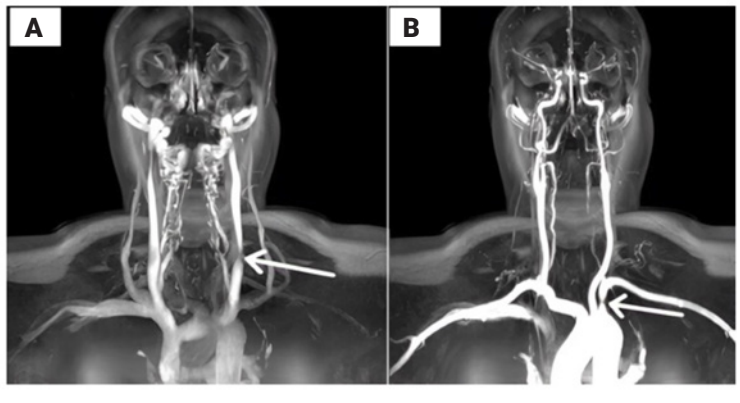

Figure 4: Chest tomography. The green arrow indicates the presence of a lesion with soft tissue density located in the inferolateral quadrant of the right breast, approximately 15 mm in size.

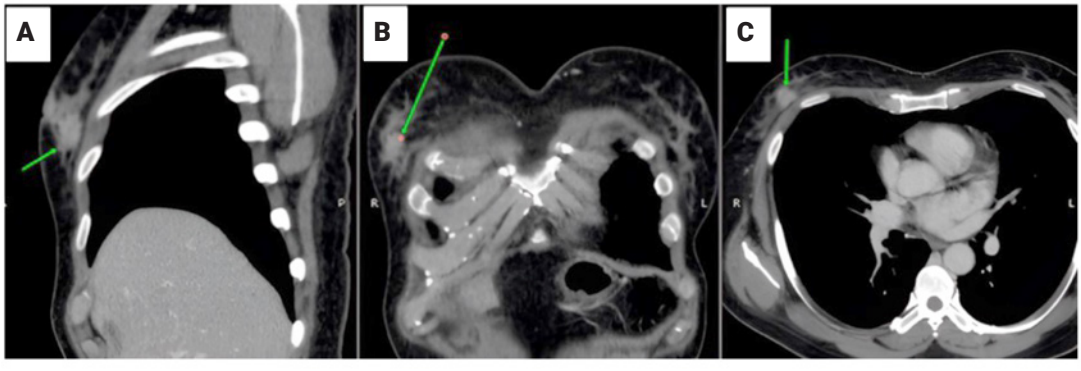

Table: Admission laboratories.

Laboratory	Result	Reference value
Hemoglobin	7.2 grams/deciliter	11-15 grams/deciliter
Hematocrit	27.3%	36-48% (woman)
Mean Corpuscular Volume	67.4 femtoliters	80-100 femtoliters
Leukocytes	18.77 x 1000/µL	4.5-11 x 1000/µL
Neutrophils	15.75 x 1000/µL	1.5-8 x 1000/µL
Platelet Count	282 x 1000/µL	150-450 x 1000/µL
Erythrocyte Sedimentation Rate	3 mm	0-15 mm
C-Reactive Protein	<0.4 milligrams/deciliter	0.4-1 milligrams/deciliter
Antinuclear Antibodies	AC-0	AC-0

DISCUSSION

The vascular manifestations of TA are variable. In more than 90% of cases, it presents with stenosis of different vascular beds, whereas aneurysmal disease occurs in approximately 25% of cases[2]. Arterial involvement affects mainly the aorta and its main branches, with a special predilection for the subclavian and common carotid arteries (left side more than right), renal, vertebral, and innominate arteries[4].

Different forms of vascular involvement are described in the literature in several types of vasculitis. For example, venous thrombosis in Behçet's disease or arterial involvement in ANCA-associated vasculitis, where the involvement of coronary and cerebral circulation can also be explained by atherosclerotic disease linked to systemic inflammation but not by vasculitic activity itself[5]. On the other hand, large vessel vasculitides, including TA, do not increase the risk of venous thromboembolic disease, but they do increase the risk of arterial involvement, similar to ANCA-associated vasculitides. In TA, an increased prevalence of cerebrovascular disease and transient ischemic attacks of 5-20% has been described and is associated with cardiovascular risk factors and atherosclerotic disease[5].

The relationship between cancer and vasculitis is rare and difficult to establish, as most reports lack the temporal relationship necessary to generate valid conclusions about this association[6]. Fain et al.[7] reported 60 cases of vasculitis associated with malignancies over a 10-year follow-up period, with a higher incidence of vasculitis in hematologic malignancies (63.1% of cases). In this group of neoplasms, the most common vasculitis is leukocytoclastic, and the only vasculitis associated with a specific neoplasm is polyarteritis nodosa with hairy cell leukemia[7]. In contrast, for TA, this association is extremely rare, and very little has been described in some cases of polycythemia rubra vera and in the context of paraneoplastic syndrome[8].

Previous research has shown that patients with TA may exhibit a hypercoagulable state, characterized by increased platelet aggregation and elevated levels of fibrinogen and D-dimer[9]. Case reports have further highlighted severe arterial thrombosis, including distal aortic and iliac involvement, emphasizing the thrombotic risk associated with this disease[10]. Genetic studies have identified associations between polymorphisms in coagulation-related genes -such as F2 G20210A, F5 G1691A, F7 G10976A, F13 G13T, FGB, ITGA2, ITGB3, and PAI-I- and the development of cardiovascular and thrombotic complications in TA patients[11]. However, while these findings suggest a potential thrombotic risk, the quality of the evidence remains moderate, and there is a clear need for studies with more robust methodologies. Given the rarity of such manifestations, it remains challenging to draw definitive conclusions regarding the relationship between TA and thrombotic events.

In clinical practice, thrombotic complications in TA can lead to diagnostic delays, particularly when the initial presentation mimics other hypercoagulable disorders. The occurrence of arterial thrombosis, especially at atypical sites such as the superior mesenteric artery, should prompt a thorough evaluation for underlying vasculitis, as well as concurrent

prothrombotic conditions, including malignancy and antiphospholipid syndrome[12]. Given this context, antiplatelet therapy has been proposed as an adjunctive measure to reduce the risk of arterial ischemic events in TA, particularly during active disease phases[13]. However, the optimal antithrombotic strategy for these patients remains to be clearly defined through prospective studies.

Our case underscores the importance of considering TA in the differential diagnosis of arterial thrombosis, particularly in young or middle-aged patients without significant atherosclerotic risk factors and highlights the need for comprehensive evaluation for potential underlying procoagulant states.

CONCLUSIONS

TA is a disease with diverse clinical presentations that can delay diagnosis. In advanced stages, vascular complications such as stenosis and atherosclerotic disease are common, predisposing patients to cerebrovascular and cardiovascular events. Unlike Behçet's disease, TA is not typically associated with thrombotic episodes, making it essential to rule out overlap with procoagulant states, including antiphospholipid syndrome and malignancy, particularly in patients with relevant risk factors.

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