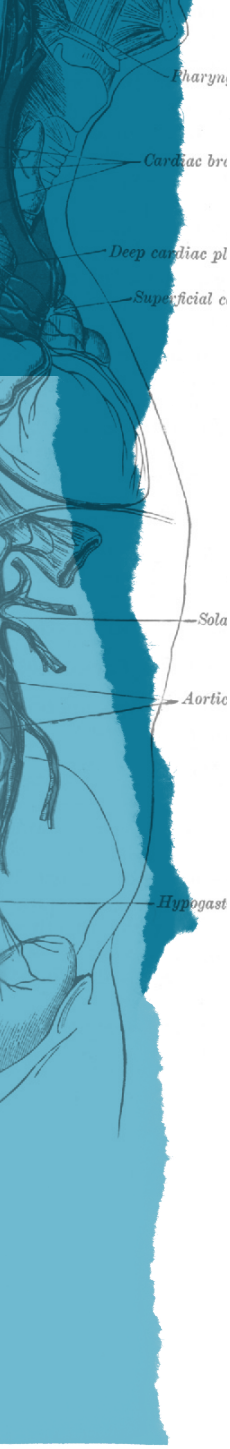

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Atypical Presentation of Giant Cell Arteritis: Case Report

Presentación atípica de arteritis de células gigantes: Reporte de caso

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ABSTRACT

Giant cell arteritis (GCA) is a granulomatous vasculitis of medium and large arteries that usually affects the aorta and/or its main branches. We report a 66-year-old, female, black, and an active smoker patient. The patient consulted due to diffuse abdominal pain, nausea, vomiting, without neurological manifestations suggestive of intracranial involvement. Vital signs within acceptable limits, pain on palpation in the epigastrium and left flank, and positive renal fist percussion. Computed tomography (CT) angiography showed intramural inflammatory lesions and Stanford type B aortic dissection; therefore, transfer to the intensive care unit was indicated. Vascular surgery suggested intramural hematoma of the descending aorta and ulcer adjacent to the minor celiac trunk. Oral beta-blocker was started. Markers and an electrocardiogram were taken without findings of acute coronary cause. Control CT angiography showed thickening of the aortic walls from the arch to the bifurcation consistent with aortitis with elevated acute phase reactants. Pain improved and the patient was transferred to the general ward. Control images indicated suspicion of GCA vasculitis, so management with corticosteroids was started. Patient reported pain again, and a magnetic resonance (MRI) angiography was requested. It showed diffuse and concentric thickening of the aortic walls from the arch to the bifurcation. This suggested an inflammatory process of the aortic wall. After 7 days of treatment with prednisolone, patient was discharged due to decreased pain and no recurrence of other symptoms. Medication was indicated to continue, and a control MRI angiography was requested. Significant pain and imaging improvement was found, so the corticosteroid dose was tapered until it was discontinued.

Keywords: Aortitis, computed tomography angiography, giant cell arteritis, magnetic resonance angiography, vasculitis.

RESUMEN

La arteritis de células gigantes (ACG) es una vasculitis granulomatosa de arterias medianas y grandes que suele afectar a la aorta y/o a sus ramas principales. Reportamos paciente de sexo femenino, 66 años, raza negra y fumadora activa. La paciente consultó por dolor abdominal difuso, náuseas, vómitos y sin manifestaciones neurológicas sugestivas de afección intracraneal. Signos vitales en límites aceptables, dolor a la palpación en epigastrio y flanco izquierdo, y puño percusión renal positiva. La angiotomografía computarizada (angio-TAC) mostró lesiones inflamatorias intramurales y disección aórtica Stanford tipo B por lo que se indicó traslado a unidad de cuidado intensivo. Cirugía vascular sugirió hematoma intramural de la aorta descendente y úlcera adyacente a tronco celíaco menor. Se inició beta bloqueador oral. Se tomaron marcadores y electrocardiograma sin encontrar

causa coronaria aguda. La angio-TAC de control mostró engrosamiento de las paredes aórticas desde el cayado hasta la bifurcación compatible con aortitis con elevación de reactantes de fase aguda. Hubo mejoría del dolor y la paciente fue trasladada a sala general. Las imágenes de control indicaron sospecha de vasculitis por ACG, por lo que se inició manejo con corticoide. La paciente refirió dolor nuevamente, y se solicitó angioresonancia magnética (angio-RM), que mostró engrosamiento difuso y concéntrico de las paredes aórticas desde el cayado hasta la bifurcación. Esto sugirió proceso inflamatorio de la pared aórtica. Luego de 7 días de tratamiento con prednisolona, se dio egreso por disminución del dolor y no recurrencia de otros síntomas. Se indicó continuar con medicamentos y angio-RM de control. La paciente presentó mejoría importante del dolor y mejoría imagenológica, por lo que se disminuyó la dosis de corticoide hasta suspenderlo.

Palabras clave: Angiografía por resonancia magnética, angiografía por tomografía computarizada, aortitis, arteritis de células gigantes, vasculitis.

INTRODUCTION

Inflammatory changes present in the aortic vascular wall are known as aortitis. It can have a broad etiological spectrum that includes infectious and non-infectious diseases, with a non-specific clinical presentation, which makes diagnosis difficult [1-3]. It is uncommon, and most cases are diagnosed with the anatomopathological study of aneurysms. The most common causes are rheumatic, and giant cell arteritis (GCA) represents more than 75% of cases [4].

GCA is a granulomatous vasculitis of medium and large arteries. It usually affects the aorta, predominantly in branches of the carotid and vertebral arteries. This condition affects the aortic wall in 15-22% of cases, and it implies a probability of suffering aortic aneurysms 17 times higher than in a normal person [4]. It classically manifests with headache, back pain, symptoms of polymyalgia rheumatica, such as pain and stiffness in the neck, shoulders, and/or pelvic girdle, and fever [5]. It can also present as severe aortic insufficiency or an aortic aneurysm.

The diagnostic criteria for GCA are: age ≥50 years, headache, abnormality of the temporal artery, erythrocyte sedimentation rate (ESR) ≥50 mm/h, and arterial biopsy with vasculitis. The presence of 3 criteria shows a sensitivity of 94% and a specificity of 91% for the diagnosis [6]. Ischemic symptoms can correlate with relapse of GCA, which can be accompanied by aortic inflammation that can progress to dilation, stenosis, or dissection (EULAR 2018) [5]. Temporal artery biopsy

is the primary method of diagnosis to establish GCA, with findings of panarteritis with mononuclear infiltrates that penetrate all layers of the arterial wall [7]; however, if there is high clinical suspicion and the imaging findings are compatible with GCA, a biopsy may not be necessary (EULAR recommendation level 2) [8].

Computed tomography angiography (CT angiography) is the first diagnostic approach due to its availability and because it allows for differential diagnosis with other causes of chest pain (Recommendation Grade C, Level 3 EULAR) [8]. However, some diseases can be confused with acute aortitis, such as aortic dissection, intramural hematoma, and penetrating atherosclerotic ulcer [9]. Magnetic resonance angiography (MR angiography) is superior and the technique of choice for diagnosis and follow-up since it allows demonstrating inflammatory activity or edema of the vascular wall and can reveal structural changes in detail [6,9] (Grade B Recommendation, Level 2B EULAR) [8]. Biopsy provides little in terms of establishing the underlying cause, considering that treatment is guided by etiology [10].

CLINICAL CASE

A 66-year-old female patient, black, from the department of La Guajira, Colombia. The patient was admitted to the emergency department with a clinical picture of 6 hours of evolution characterized by sharp and sudden retrosternal pain radiating to the lumbar region, nausea, vomiting, and loss of consciousness. She reported a history of cholecystectomy and active smoking. Vital signs showed: blood pressure = 140/70 mmHg, mean ventricular rate = 81 beats/minute, respiratory rate = 18 breaths/minute, temperature = 36.5°C, pain analog scale = 10/10, and scale Glasgow = 15/15. The pain was found in the left flank and epigastrium, positive left fist percussion, diaphoresis, and paresthesia in the lower limbs. Normal troponin curve with an electrocardiogram (ECG) without changes.

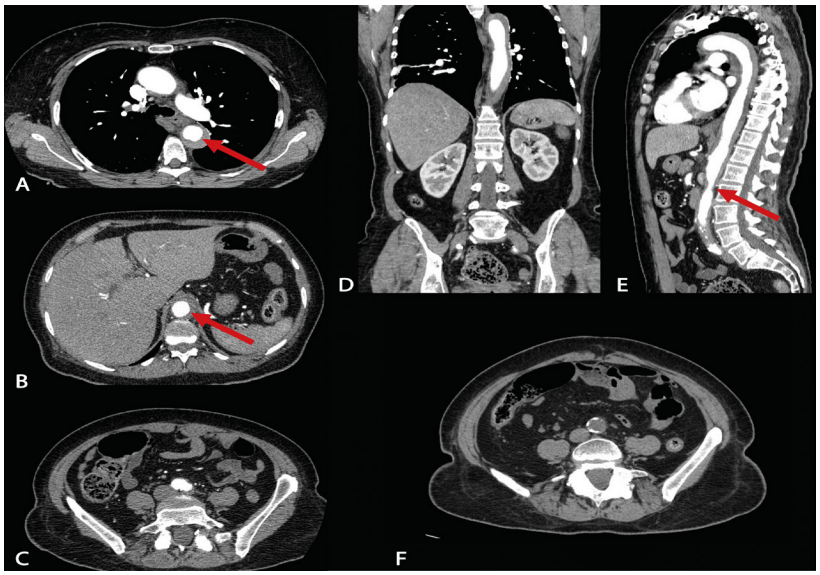
Laboratory tests showed negative troponins and decreased hemoglobin and leukocytes = 11.07 × 109/L (table). An echocardiogram and CT angiography of the abdomen and chest were requested due to the high probability of aortic dissection. CT angiography showed Stanford type B aortic dissection and flap from the aortic arch at the level of the middle third (figure 1). Aortic dissection was diagnosed, and vascular surgery was consulted. The echocardiogram showed no abnor-

malities, and management with labetalol was started. The patient was reassessed, labetalol was discontinued, and management with metoprolol was started.

Table. Alterations in Paraclinical Tests of the Reported Case

Date	Test	Result	Parameter
28/09/2020	Leukocytes	13,88	4,50-11,30
30/09/2020	Glycemia	112,70 mg/dL	70-100
04/10/2020	Rheumatoid Factor	22,50 UI/mL	0-14
	C-Reactive Protein	328,81 mg/l	0-5
	C3 Complement Fraction	1,92 g/L	0,9-1,8
	C4 Complement Fraction	0,42 g/L	
	Antinuclear antibodies	Negative	
	VSG	81	

Source: own elaboration.



Note. Slightly isodense, concentric, and circumferential (6-9 mm) wall thickening from the middle third of the aortic arch to the iliac bifurcation, associated with altered fat density in the posterior mediastinum and retroperitoneum (AE). Mild to moderate calcified aortic atheromatosis, especially in the distal abdominal aorta and iliac arteries (F).

Source: own elaboration (based on information obtained from medical records).

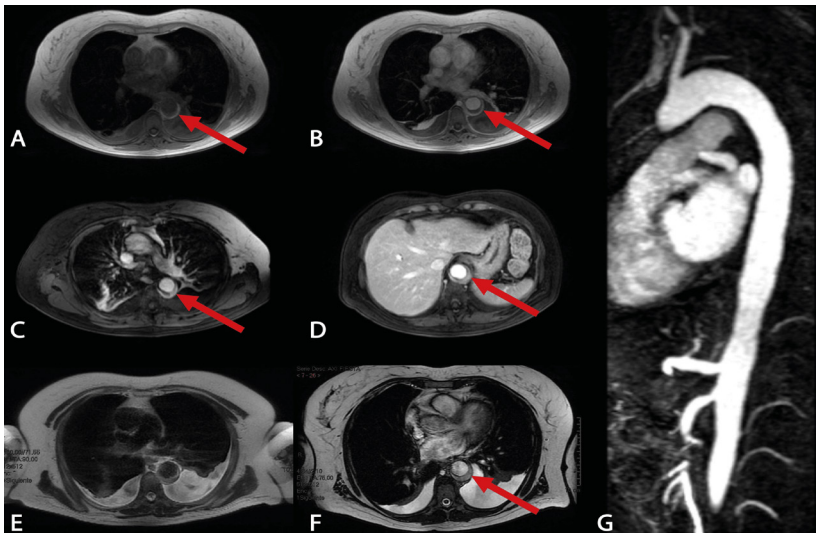
Figure 1. CT Angiography

Subsequently, the patient was transferred to the intensive care unit (ICU). Cardiovascular surgery evaluated CT angiography and ruled out aortic dissection. Intramural hematoma of the descending aorta with an ulcer adjacent to the celiac trunk (<1 cm) was determined, without requiring surgical correction (figure 1). During the ICU stay, the patient reported pain of greater intensity in the interscapular region associated with oxygen desaturation (89%). There were no electrocardiographic changes or troponin elevation. Table 1 illustrates the alterations in paraclinical tests of the reported case.

One day later, there was an improvement in the clinical picture without vasopressor support. However, she was not transferred to the general ward due to the persistence of atypical subscapular pain and desaturation. Control CT angiography was ordered, which showed thickening of the aortic walls from the arch to the bifurcation, compatible with aortitis. Surgical management was ruled out.

The patient was transferred to the general ward with stable hemodynamic conditions and pain improvement. Internal medicine indicated suspicion of aortitis associated with vasculitis, and due to the patient's age, it suggested possible GCA. Additionally, acute phase reactants were requested. Management with corticosteroids was started due to clinical suspicion of autoimmune aortitis.

Contrast-enhanced MR angiography of the chest and abdomen showed aortic atheromatosis, and diffuse and concentric thickening of the aortic walls from the arch to the bifurcation (figure 2). Contrast medium enhancement suggested an inflammatory process of the aortic wall (figure 2). The patient presented an improvement in her clinical condition with hemodynamic stability and decreased precordial pain. She was discharged with an indication of prednisolone 50mg/day for 8 weeks, esomeprazole, alprazolam, atorvastatin, metoprolol, quetiapine, bisacodyl, and pregabalin. Angio-MR of the abdomen (control) and outpatient control with internal medicine and rheumatology were indicated.



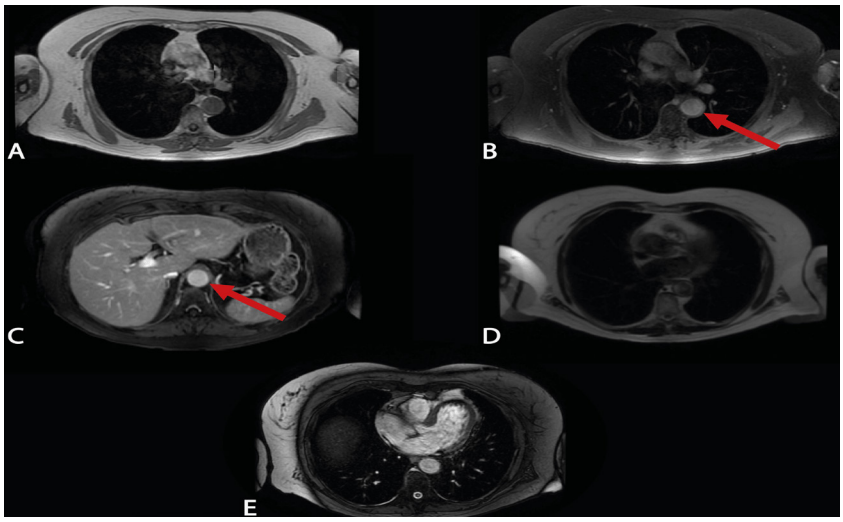
Note. Concentric enhancement of the thoracic and abdominal aortic walls in B, C and D (with contrast medium), with respect to A (without contrast). Diffuse para-aortic hyperintensity in T2- and Fiesta-weighted sequences (6-9 mm) corresponding to the vascular inflammatory process (E and F). Symmetric opacification without evidence of dissection flaps or hematomas (G).

Source: own elaboration (based on information obtained from medical records).

Figure 2. Magnetic Resonance Angiography of the Thorax and Abdomen

During the first control, significant clinical improvement and stable evolution were observed. Corticosteroid taper was initiated, and additional MR angiography was requested. In the second control appointment, improvement in the clinical picture was observed. The MR angiography report showed an almost complete disappearance of the thickening of the thoracic and abdominal aorta walls (figure 3), which is consistent with an inflammatory process. Aortitis resolved, and corticosteroid management was discontinued.

Being a patient older than 50 years, with ESR >50 mm/h, elevated C-reactive protein, radiological findings (described above), resolution of the condition with corticosteroids, and a positive rheumatoid profile, the diagnosis of GCA was confirmed. However, no biopsy sample was obtained under the American College of Rheumatology (ACR) classification criteria.



Note. Almost complete disappearance of aortic wall thickening from the middle third of the arch to the iliac bifurcation. Mediastinal and retroperitoneal fat signal is preserved.

Source: own elaboration (based on information obtained from medical records).

Figure 3. Magnetic Resonance Angiography (30 days after treatment)

Continúa...

DISCUSSION

As strengths in the case management, we find the disposition of radiological resources and specialized laboratories, and the ability of professionals to identify significant radiological findings, correlate them with clinical symptoms, and consider the diagnosis of GCA, taking into account that it is not common. According to the EULAR, with a level 1 recommendation, early management with diagnostic images is recommended to complement GCA diagnostic criteria [5,8]. Regarding the limitations, we found the atypical presentation of the condition and the non-performance of a biopsy to confirm the diagnosis due to the risks and possible complications. It is important to note that for EULAR with a level 2 recommendation, patients with a positive imaging test and high suspicion of GCA do not need further tests to confirm the diagnosis, so in our case, the biopsy was not mandatory [8].

As there is no specific clinical biomarker to control the disease activity and considering the evaluation of laboratory findings is difficult, several working groups, such as the one of A. Habouchi has proposed identifying radiological changes in the anterior mediastinal fat during the disease and its probable predictive value with active inflammatory disease. Based on this, they have found that patients with high density (>10UH) have active disease [11], a fact that was evidenced in our patient.

Thoracic pain is a common symptom in the emergency room and deserves special attention due to the numerous diagnostic possibilities. An adequate history, physical examination, and complementary tests are essential for the differential diagnosis. In the anamnesis the patient did not express and denied cranial symptoms such as headache, visual disturbances, mandibular and/or lingual claudication, morning stiffness in shoulders and/or neck. Physical examination revealed no neurological symptoms such as eyelid ptosis, preserved pupillary reflexes, no visual acuity alterations, no ophthalmoplegia, no scalp allodynia and no pain on examination of the temporal arteries [14]. The diagnosis of acute coronary syndrome was eliminated as no coronary obstructions could be identified on imaging, and the ECG showed no signs of myocardial ischemia. However, changes in CT angiography performed to investigate acute aortic syndrome and aortic dissection raised the diagnostic hypothesis of aortitis and could be related to disease relapse [12].

In our case, the clinical picture was compatible with acute aortic syndrome (as evidenced by the images), and the symptoms indicated acute aortitis of autoimmune etiology. Comparing the 1990 ACG criteria with the updated 2022 criteria, our patient does not apply to think of such diagnosis, since she is older than 50 years, but has a laboratory criterion (ESR >50mm/h or CRP >10mg/l) which is not enough score to consider giant cell arteritis although for the 1990 criteria she would fulfill (female sex, age >50 years and ESR >50 mm/h). [13, 14]. However, in this case, the patient had no neurological symptoms or signs suggestive of intracranial vessel involvement, so the initial approach was to search for a thoracoabdominal cause. Therefore, taking into account the clinical and imaging features and the resolution of the case with the use of steroids, the autoimmune etiology was demonstrated. According to the above, it is important to mention that reports of extracranial involvement have been found, as in this case [15]. In extracranial cases there is an important relationship between the findings of Doppler ultrasound of axillary arteries and systemic arterial involvement. However, in our institution we do not have the appropriate personnel to perform it [16].

CONCLUSIONS

In managing this type of case, it is essential to investigate and delve into each clinical aspect of the patient, relate the imaging findings with the clinical ones, and not rule out a vascular inflammatory cause.

Considering that not all acute aortic syndrome is an aortic dissection, aortitis should be regarded as a possibility, and early diagnosis and treatment can change the prognosis.

We emphasize that when presenting with a clinical picture of acute aortic syndrome, vasculitis should be considered as a differential diagnosis.

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