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Case report

Unstable angina secondary to left main coronary artery vasculitis as a rare initial manifestation of systemic lupus erythematosus. A case report

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ABSTRACT

We present the case of a woman in her third decade of life, known to have difficult-to-control mixed headaches and polycystic ovary syndrome, under hormonal treatment. Without any other manifestation, the patient debuted with an acute coronary syndrome classified as unstable angina. Electrocardiogram showed anterior and lateral ST segment depression and ST elevation in aVR. Coronary computer tomography and coronary angiography showed evidence of significant obstruction of the left main coronary artery. The patient was diagnosed with systemic lupus erythematosus (SLE), and was classified as vasculitis secondary to SLE as an unusual initial manifestation.

Keywords: Lupus Erythematosus, Systemic; Vasculitis; Anomalous Left Coronary Artery; Unstable, Angina (Source: MeSH - NLM).

RESUMEN

Angina inestable secundaria a vasculitis del tronco coronario izquierdo como manifestación inicial poco frecuente de lupus eritematoso sistémico. Reporte de un caso

Se presenta el caso de una mujer en la tercera década de la vida, con cefalea mixta de difícil control y síndrome de ovario poliquístico bajo tratamiento hormonal, sin ninguna otra manifestación, la cual debutó con un síndrome coronario agudo tipo angina inestable. El electrocardiograma mostró infradesnivel en cara anterior y lateral, así como supradesnivel del ST en aVR. La angiotomografia de coronarias y la coronariografía mostraron evidencia de obstrucción importante del tronco de la coronaria izquierda. Tras el abordaje se llegó al diagnóstico de vasculitis secundaria a lupus eritematoso sistémico como manifestación inicial poco habitual.

Palabras clave: Lupus Eritematoso Sistémico; Vasculitis; Arteria Coronaria Izquierda Anómala; Angina Inestable (Fuente: DeCS - Bireme).

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease that can affect any part of the body, resulting in a wide range of clinical presentations. Within its manifestations, vasculitis occurs in 11 to 36% of patients, especially in disease outbreaks [1,2].
In general, cutaneous vasculitis represents the most common form of presentation in up to 69% of cases [2].

On the other hand, coronary artery vasculitis is one of the rarest conditions, with only a few cases reported in the literature, and is usually suggestive of a severe outbreak of the disease. It can be accompanied by fever, fatigue, anemia, increased acute phase reactants, livedo-reticularis, and weight loss [2]. However, it can manifest in the absence of a clinical SLE outbreak or laboratory evidence of disease activity [3].

We present the unusual presentation of a young woman whose initial manifestation of SLE was an acute coronary syndrome secondary to vasculitis of the main left coronary artery.

Case report

This is a 24-year-old female patient, with previous history of difficult-to-control mixed headache (tensional and migraine), polycystic ovarian syndrome under hormonal treatment (progestin), and non-painful oral ulcers.

She had emergency care due to oppressive chest pain (9/10 intensity), lasting approximately 120 minutes, without radiation or evidence of adrenergic discharge. During her evaluation, a 12-lead electrocardiogram showed ST segment depression in the anterior, upper and lower lateral leads, as well as ST segment elevation in aVR lead. **(Figure 1A)**. Laboratory biomarkers of myocardial damage were normal (troponine I: 0.71 ng/ml, second determination 0.1 ng/ml (normal value: 0-1 ng/ml)), fulfilling the criteria for high-risk non-ST elevation acute coronary syndrome (ACS) due to electrocardiographic changes, highly suggestive of left main coronary artery involvement. The need for coronary angiography with the possibility of

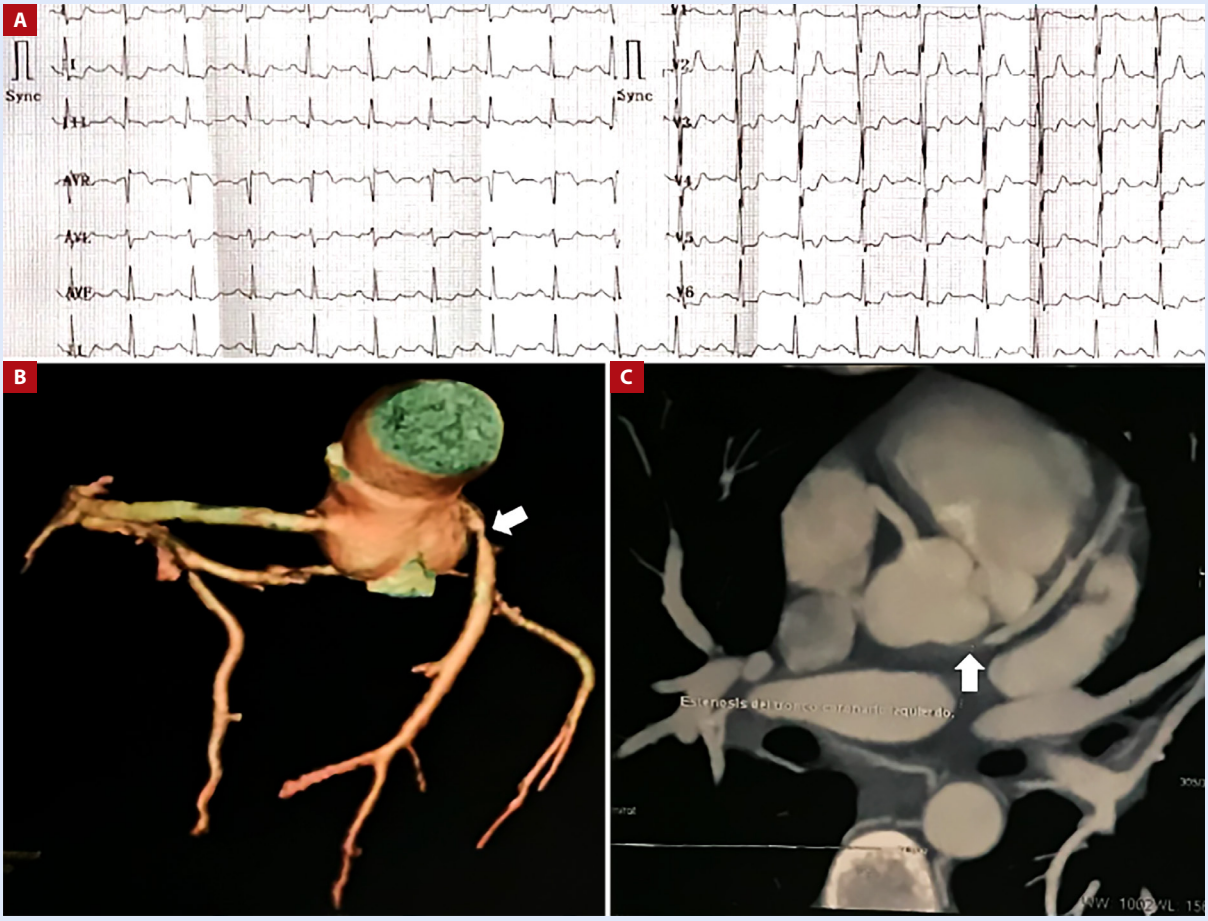

Figure 1. A) Twelve-lead electrocardiogram on admission to the Emergency Department. ST-segment depression can be seen in V2-V6 and DI-aVL leads, as well as ST-segment elevation in aVR. **B)** 3D reconstruction, and **C)** Axial view of angio tomography of coronary arteries. Significant stenosis is observed at the level of the main left coronary artery.

angioplasty was discussed, but the patient initially decided not to accept the procedure. Therefore, coronary computer tomography (CCT) was performed, revealing 90% obstruction of the left main coronary artery **(Figures 1B and 1C)**. Anti-ischemic management was provided (acetylsalicylic acid 300 mg, clopidogrel 300 mg, enoxaparin at anticoagulation doses, and atorvastatin) and unstable angina was diagnosed in a patient without apparent cardiovascular risk factors.

Initially, the patient did not agree to undergo coronary angiography, so an etiological approach was initiated. In the context of a young woman without clear cardiovascular risk factors, antinuclear antibodies (ANAs) were requested by indirect immunofluorescence (IIF) with 1:320 dilution and fine speckled pattern. We found positive anti-dsDNA (double-strand DNA) antibodies, erythrocyte sedimentation rate (ESR) at 33 mm/1 hour, C-reactive protein (PCR) at 27 mg/ L, leucopenia at $4x10^3/mm^3$, as well as previous history of non-painful oral ulcers, meeting criteria for SLE (11 points) according to the American College of Rheumatology / European League Against Rheumatism (ACR/EULAR) – 2019 criteria. In **Table 1**, we provide the complete immunological panel of the patient.

Prednisone 50 mg QD and mycophenolic acid 500 mg BID were established, and coronary angiography was performed once the acute phase reactants decreased after one month. Before coronary angiography, transthoracic echocardiography showed preserved left ventricular ejection fraction (70%), without alterations in mobility or segmental thickening at rest, and without significant valve disease. Coronary angiography showed obstruction of 95% of the left main coronary artery in different projections **(Figures 2A and 2C)**, so a drug-eluting stent was placed successfully **(Figures 2B and 2D)**. Its proper placement was verified by intravascular ultrasound (IVUS) without detecting other added lesions.

The patient was discharged with rivaroxaban-based oral anticoagulation (3 months), double antiplatelet therapy (one year), and high-intensity statin therapy. In the follow-up of the patient two years after the episode, she continued with acetylsalicylic acid 100 mg QD and atorvastatin 40 mg QD, mycophenolic acid 500 mg BID, and hydroxychloroquine 200 mg BID. The steroid is currently suspended without new episodes of angina or other outbreaks.

Discussion

Cardiovascular disease is a common manifestation of SLE and continues to be one of the leading causes of death in these patients. Various studies have shown a process of "accelerated atherosclerosis," which increases the risk of cardiovascular events [4]. In this sense, Tektonidou *et al.* [5] described that the rates of hospitalization for acute myocardial infarction and ischemic stroke increased over time in patients with SLE in both younger and older women, while the rates of unstable angina decreased (probably due to the advent of high-sensitivity troponin).

According to a national data base from 2010 to 2019 in the United States, a total of 7,076,477 ACS hospitalizations were identified, of which 2.1% were related to coronary vasculitis. Among the main causes, SLE was reported in 4.1/1000 cases [6]. Vasculitis is a rare manifestation of SLE, with coronary artery disease being one of the least frequent. There are only a few cases reported in the literature. For example, a case of a 22-year-old patient who, after 4 years of diagnosis of SLE, presented an ST-segment elevation myocardial infarction, despite adequate treatment of the disease [7]. Contrary to our case, the patient had no other manifestation associated with SLE.

Table 1. Complementary laboratory findings during clinical approach

Laboratory	Result	Reference values
Prothrombin time (seconds)	14	14
Partial Activated Thromboplastin Time (seconds)	30	34
Rheumatoid factor (UI/mL)	7.5	0.00 - 15.00
Lupus anticoagulant	Negative	Negative
Anti-B2 glycoprotein 1 antibodies (U.G)	3.3	0.0 - 20
Anti-cardiolipin IgG antibodies (GLP/mL)	5.6	0.0 - 11.0
Anti-cardiolipin IgM antibodies (MPL/mL)	6	0.0 - 11.0
Anti-SCL70 antibodies (U/mL)	17	0.0 - 25.0
Anti-SM antibodies	Negative	Negative

GPL: amount in micrograms of the antibodies in a determinate volume of serum; MPL: amount in micrograms of the antibodies in a determinate volume of serum; U.G: micrograms.

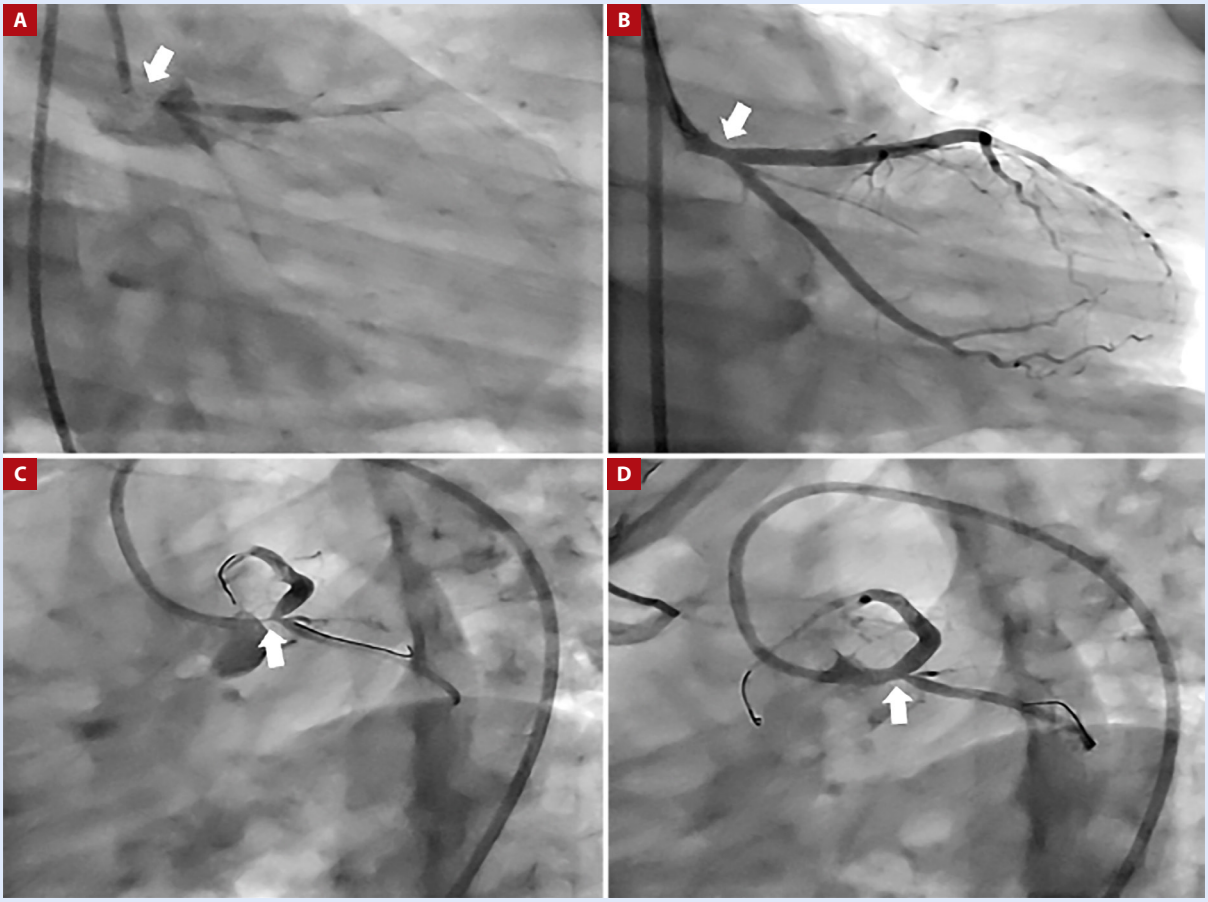

Figure 2. A) Cranial antero-posterior projection. The white arrow indicates practically total obstruction of the main left coronary artery, with minimal passage of contrast. **B)** The white arrow indicates the occlusion site after placement of a drug-eluting stent, with recovery of caliber and flow. **C)** Caudal left anterior oblique projection (spider view). The white arrow indicates an almost total occlusion of the left main coronary artery. **D)** The white arrow shows significant improvement of caliber and flow after placement of the drug-eluting stent.

In a retrospective cohort, it was reported that of 565 patients diagnosed with SLE, 191 (33.4%) developed vasculitis, of which cutaneous vasculitis was the most frequent, reported in 60% of patients. Regarding isolated coronary vasculitis, only one case (0.5%) was reported and only one other case in combination with skin compromise. In addition, patients who developed vasculitis had other cardiovascular manifestations such as livedo reticularis, Raynaud's phenomenon, digital gangrene, thrombosis, and cranial neuropathy, as well as more advanced and juvenile-onset disease [8]. This is consistent with our patient's clinic and with a retrospective cohort of patients with juvenile-onset SLE in Taiwan over a 20-year period by Smith *et al*., who reported that 6 of 157 patients exhibited angiographically demonstrated coronary abnormalities (dilation, aneurysms, vasculitis, and stenosis) [9].

Regarding in-hospital outcomes in patients with ACS and SLE, De Matos-Soeiro and colleagues [10] described that 91% were women (eleven patients), with a median age of 47 years. Most patients presented typical precordial pain (91%), being the anterior descending artery the most often affected vessel (73%). One patient underwent coronary artery bypass grafting, seven underwent percutaneous coronary intervention with bare-metal stents, and three were treated medically with an in-hospital mortality rate of 18%. In a Mexican cohort, Zonana-Nacach *et al.* [11] described that of 41 patients with severe SLE outbreak, six patients presented with vasculitis and one fatal case of acute myocardial infarction. This data reveals the low prevalence of ACS related to coronary vasculitis in patients with SLE in our population.

Diagnosis can be made using non-invasive studies such as CCT or direct visualization using coronary angiography [12]. Saparia *et al.* described a case of coronary vasculitis in a 47-year-old woman with lupus nephritis; coronary angiography revealed multiple saccular lesions and irregularities in the trajectory of both coronary arteries. Fifteen months after the establishment of immunosuppressive management, a new coronary angiography was performed, finding resolution of practically all the lesions [13].

Due to the extremely low frequency of this manifestation, there is no consensus regarding its treatment. Smith *et al.* suggest management with corticosteroids and cyclophosphamide [9]. To the best of our knowledge, this is the first case managed with stent placement due to significant obstruction of the left main coronary artery. So, it is extremely important to report this type of case and observe its evolution.

Finally, coronary artery vasculitis is an unusual presentation of SLE and even more so as the initial presentation of the disease. In young patients (mainly women) rheumatic diseases should be considered as a cause of ACS.

Ethical issues

Unfortunately, because the case was obtained retrospectively, it was not possible to obtain informed consent. In addition, due to this situation, it was not submitted to the ethics committee, highlighting that at no time the identity of the patient was revealed

Author´s contributions

MAPG: Conceptualization and writing original draft; JPG: Data curation and conceptualization; **JAGD:** Writing, review and editing; **JJAP:** Writing original draft; **CYSU:** Investigation; JPVP: Supervision and project administration.

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