CLINICAL CASE
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Mesenteric ischemia as a presentation of primary antiphospholipid syndrome in a male patient

Isquemia mesentérica como presentación de síndrome antifosfolípido primario en paciente masculino

Erick Zúñiga-Garza,* Francisco Franco-López‡

Keywords:
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Palabras clave:
isquemia mesentérica, síndrome de anticuerpos antifosfolípidos, abdomen agudo.

ABSTRACT

Antiphospholipid syndrome is an autoimmune disease characterized by the thromboembolic formation and maternal morbidity, associated with a persistent increase in antiphospholipid antibody titers; involvement in male patients is rare, and onset with gastrointestinal manifestations is uncommon; for this reason, we present the clinical case of a 19-year-old male patient with no relevant history, who started with antiphospholipid syndrome through mesenteric ischemia.

RESUMEN

El síndrome antifosfolípido es una enfermedad autoinmune caracterizada por formación tromboembólica y/o morbilidad materna, asociada a incremento persistente en los títulos de anticuerpos antifosfolípidos, la afectación en pacientes masculinos es rara, y el inicio con manifestaciones gastrointestinales es algo poco común, por dicha razón presentamos el caso clínico de paciente masculino de 19 años sin antecedentes de importancia, quien inicia con un síndrome antifosfolípido a través de una isquemia mesentérica.

INTRODUCTION

Antiphospholipid syndrome (APS) is a multisystem disease characterized by thrombus formation and emboli associated with maternal morbidity and persistent increase in antiphospholipid antibody titers;[1] 1% of cases present as catastrophic APS, in which multiple vascular occlusive events occur affecting perfusion of various organs.[2] A small percentage of patients present with gastrointestinal involvement.[3]

CASE PRESENTATION

A 19-year-old male with no previous history came to the emergency department for two weeks of intermittent abdominal pain located in the epigastrium with irradiation towards the mesogastrium, described as oppressive, only referred to a change in bowel habits, previously treated with proton pump inhibitor and butylhyoscine/lysine as analgesic without improvement. He went twice to the emergency department with the same symptoms; on each visit he was treated conservatively. On his fourth visit, he mentioned that pain had increased and was accompanied by vomiting. Physical examination revealed dehydrated mucous membranes and generalized pallor, tachycardia (100 bpm), and abdomen with pain on palpation in the epigastrium, with evidence of peritoneal irritation. H was admitted for further diagnostic protocol with the following paraclinical tests:

Urea 40.3 mg/dl, creatinine 1.99 mg/dl, sodium 139 mEq/l, potassium 3.6 mEq/l, chlorine 104 mEq/l, white blood cells 15,070

* Resident of general surgery.
‡ Attending physician, General Surgery Service. Centennial Hospital Miguel Hidalgo.

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10^3/l, neutrophils 65%, hemoglobin 15.7 g/dl, hematocrit 47%, platelets 370,000 10^3/l, prothrombin time 15.1 seconds, thromboplastin time 25.2 seconds, INR 1.2.

The abdominal ultrasound showed free fluid in subhepatic, peri splenic, and para vesical recesses *(Figure 1)*.

Due to these findings, an exploratory laparotomy was performed where abundant inflammatory response fluid (900 ml), small bowel loops (120 cm) with thrombosis of mesenteric vessels were seen. An ischemic segment resection with Brooke ileostomy at 150 cm from the angle of Treitz and Hartmann closure of the distal stump *(Figure 2)* were performed. It was decided to place a Bogota bag for a *second look* laparotomy.

Subsequently, he was sent to the general surgery floor, where he presented an adequate stoma and Bogota pouch expense. An evaluation by the internal medicine, hematology, and rheumatology services was requested, who suggested taking anticardiolipin antibodies, anti-Beta-2-glycoprotein antibodies, antinuclear antibodies, and homocysteine levels. Treatment was started with enoxaparin, then with acenocoumarin.

After 48 hours he was reoperated (second look). The Bogota bag ass withdrawn. A few thrombosed vessels on the non-ischemic omentum were found. The small bowel loops were dilated, edematous, and without peristalsis. The mesentery was thickened, cramped, with multiple swollen lymph nodes. The distal segment (stoma) had no signs of ischemia. The terminal ileum stump showed no leak *(Figure 3)*. The abdominal wall was closed with a continuous suture with polydioxanone 1 in two stages, and Blanco Benavides stitches were used with braided polyester 5.

Subsequently, he continued his evolution on the general surgery floor, where the suggested laboratory results were obtained (Ac anticardiolipin IgG 59, IgM 50.2), and a diagnosis of primary antiphospholipid syndrome was made. The patient was discharged with a functional ileostomy. Follow-up by general outpatient surgery and rheumatology continued, and intestinal reconnection was performed electively at six months *(Figure 4)* without complications.

DISCUSSION

Between 1983 and 1986, a clinical syndrome of thrombosis associated with antibodies against phospholipids was identified. Initially it was named anticardiolipin syndrome and currently is known as antiphospholipid syndrome.[3] It is a multisystem disease characterized by thromboembolic formation and maternal morbidity, associated with a persistent increase in antiphospholipid antibody titers.[1] In 2019, an incidence of antiphospholipid syndrome of two cases/100,000 population was reported, with a prevalence of 50 cases/100,000 population;[4] however, no data were found in the Mexican literature search.

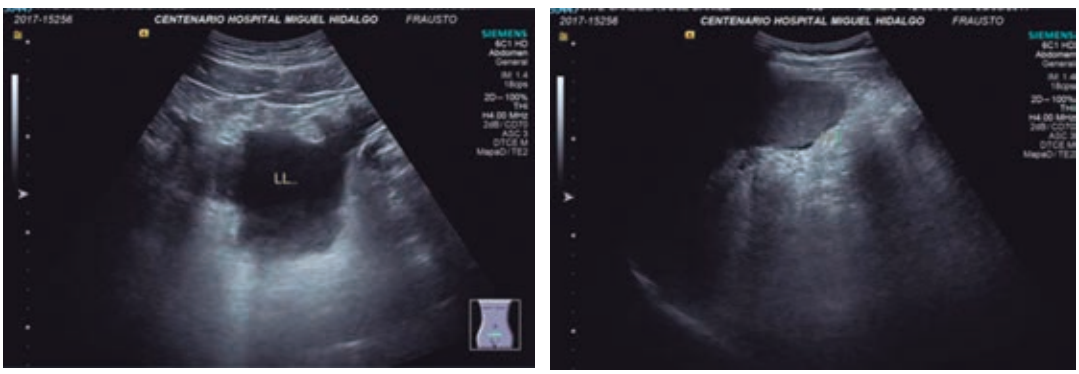

Figure 1: *Ultrasound scan showing free fluid.*

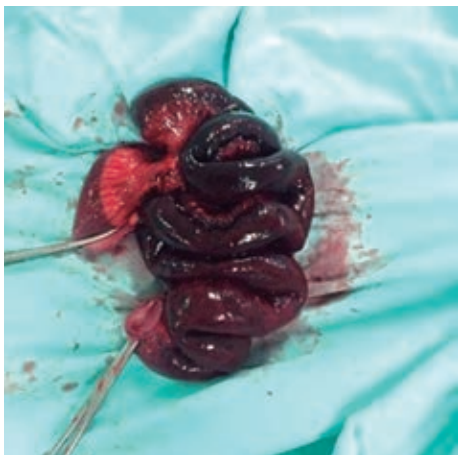

Figure 2: *The macroscopic specimen of the surgical event shows a necrotic small bowel. Histopathology reported a small bowel segment of 112 cm, with transverse dimensions of 3 × 3 cm. Microscopically there were recent multifocal arterial thrombosis and extensive pan mural ischemic necrosis.*

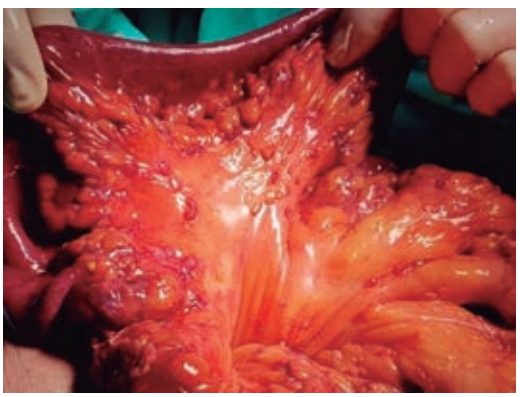

Figure 3: *Surgery (second look) showing few thrombosed vessels in the omentum, without omentum ischemia, and edematous small bowel loops dilated.*

The diagnosis of APS is made through clinical and laboratory criteria (Sapporo criteria).[5] They were updated in 2006;[6] the clinical criteria include an episode of vascular thrombosis or morbidity during pregnancy. In contrast, laboratory criteria include elevated lupus anticoagulant titers, anticardiolipin antibodies, and anti-β_2 glycoprotein antibodies.[6]

Intestinal involvement by APS is rare and is usually associated with a poor outcome.[7] Intestinal involvement may manifest as mesenteric ischemia, characterized by hypoxia of the bowel due to a sharp decrease in blood perfusion caused by embolism or thrombosis.[8]

The clinical presentation is nonspecific. Symptoms and other findings that may suggest mesenteric thrombosis are abdominal pain, diarrhea, vomiting, blood in stool, hyperlactatemia, leukocytosis, and metabolic acidosis.[8]

There is no standardized treatment for catastrophic APS, but anticoagulation is the mainstay therapy, and in some cases, surgery is necessary.[7,9] The prognosis is generally poor due to low clinical suspicion.[1,6-9]

CONCLUSION

The onset of an antiphospholipid syndrome as mesenteric ischemia is an extremely rare pathology. In a previously healthy patient, this disease should always be suspected in the range of differential diagnoses. Early recognition and multidisciplinary treatment can change the outcome of these patients.

Figure 4: *Postoperative status of the patient, showing a healed wound.*

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Correspondence:
Erick Zúñiga-Garza
E-mail: erickzunigagarza5@gmail.com