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Case Report

Catastrophic antiphospholipid antibody syndrome presenting as acute vascular occlusion in a young female patient

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ABSTRACT

Acquired thrombotic and thromboembolic disorders may be presented initially with symptoms and signs of acute ischaemia or organ dysfunction that will lead many of these patients to seek care in the emergency department. We report a case of a 19-year-old female patient who developed catastrophic antiphospholipid syndrome (CAPS syndrome or Asherson syndrome) 6 weeks post stillbirth with an initial presentation of acute vascular occlusion. The patient was immediately operated and anticoagulated with significant improvement.

Síndrome de anticuerpos antifosfolípidos catastrófico que se presenta con oclusión vascular aguda en una paciente joven

Palabras clave:
Síndrome antifosfolípido
Síndrome de Asherson
Síndrome antifosfolípido catastrófico

RESUMEN

Los trastornos trombóticos y tromboembólicos adquiridos pueden manifestarse inicialmente con signos y síntomas de isquemia aguda o disfunción orgánica que derivará a muchos de estos pacientes al servicio de urgencias. Se presenta el caso de una paciente de 19 años de edad que desarrolló un síndrome antifosfolípido catastrófico (o síndrome de Asherson) 6 semanas después del parto de un feto muerto con una presentación inicial de oclusión vascular aguda. La paciente fue intervenida inmediatamente y se inició un tratamiento con anticoagulantes que supuso una mejora significativa.

Introduction

In the 1980s and 1990s, some case reports emerged in the literature documenting patients with thrombotic complications, often fatal, associated with the presence of antiphospholipid antibodies. Only in 1992 for the first time, 10 patients with this unusual condition were revisited. The main characteristics of these were properly review in an attempt to define the severity and rapidity of installation and a catastrophic adjective was added to represent this variant of the antiphospholipid syndrome (APS).[1] Since then, the term catastrophic antiphospholipid syndrome (CAPS syndrome or Asherson syndrome) was accepted and has been used extensively.

Case description

We present a case of a 19-year-old black woman that presented to the Emergency Department with abdominal and left thigh pain. Previous medical and obstetric history included a recent stillbirth

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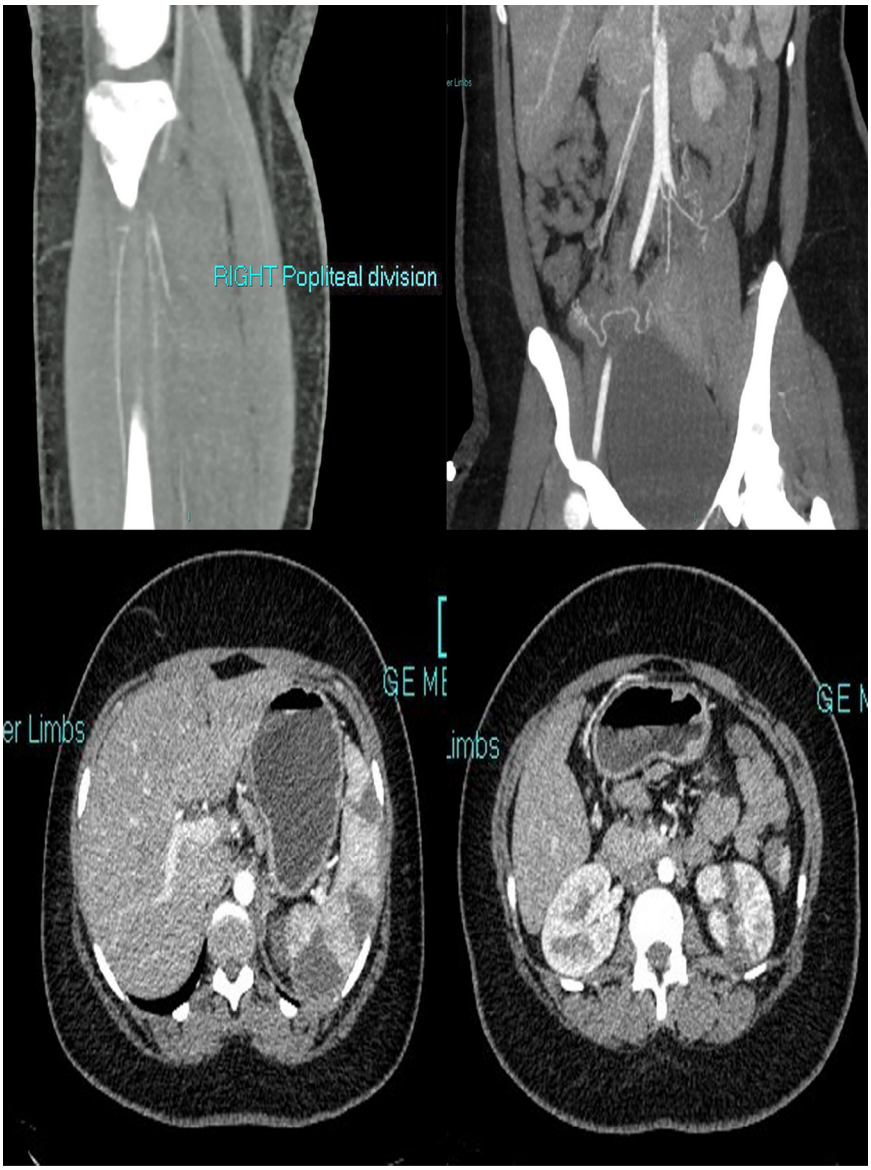

Fig. 1. Extensive fresh thrombus is noted in the left inflow, left CFA and the left SFA. Thrombus also noted in the right internal iliac, right profunda and SFA origin. Fresh thrombus noted in the right distal popliteal and trifurcation origin. Multiple splenic and left renal infarcts noted.

6 weeks ago, at 36 weeks with positive lupus anticoagulant. Recent admission to Hospital due to abdominal pain, seen by Medical and Surgical team that ruled out acute pathology and was referred to Obstetrics service that discharged the patient with further follow-up. At arrival the patient was agitated and uncooperative complaining of abdominal and left thigh pain. The patient reported severe pain in her left leg accompanied by nausea, she did not have chest pain, shortness of breath, fever, chills, dysuria, constipation, or diarrhoea. The temperature was 37.1 °C, the pulse was 91, and the respiratory rate 18. The blood pressure was 135/85 mmHg. The patient was intermittently agitated. Examination revealed diffusely tender abdomen but not distended; the bowel sounds were normal. Examination of the extremities demonstrated absent pedal pulses on both sides, and a mild sensory loss on the left leg. Morphine sulphate, paracetamol and fluids were administered. A point-of-care ultrasound of lower legs showed the absence of a flow signal suggestive of vascular occlusion.

Initially the patient was suspected to have a sickle cell pain crisis however a reviewed of recent blood tests including electrophoresis did not show the presence of homozygous HbS or documented other hemoglobinopathies. Blood tests revealed; Hb 110 g/L, WBC 13.6×10^9/L, Platelets 151×10^9/L, total bilirubin 5 μmol/L, ALT 30 U/L, GGT 98 U/L, calcium 2.36 mmol/L, phosphate 0.95 mmol/L, ALP 125 U/L, sodium 136 mmol/L, potassium 3.4 mmol/L, urea 3.0 mmol/L, creatinine 83 μmol/L (reference range: 0–110), AKI Stage 1, blood gases showed lactic acidosis type A. Increased reticulocytes and free haemoglobin indicated a hemolytic anaemia with scanty schistocytes on peripheral blood smear analysis. Troponin T 61, (reference range: <14) with no ST segment changes on electrocardiography.

An emergency CT (Fig. 1) was requested that reported extensive fresh thrombus is noted in the left inflow, left common femoral artery and the left superficial femoral artery. Thrombus also noted in the right internal iliac, right profound and superficial femoral artery origin. Fresh thrombus noted in the right distal popliteal and trifurcation origin. Multiple splenic and left renal infarcts noted, a CT brain did not reported acute ischaemic changes. The trachea was intubated, and ventilatory assistance was begun. Vascular surgeons were requested and the patient was operated on: under general anaesthesia, a left iliofemoral embolectomy associated to a