Clinical Case Reports International

Case Report
Published: 14 Sep, 2022

Probable CAPS after Surgical Intervention in a Patient with Known Ulcerative Colitis: A Case Report and Literature Review

Sarah Assaf, Ralph Daher, Rodrigue Chemali and Fadi Hayek*

Lebanese American University, Lebanon

Abstract

Catastrophic Antiphospholipid Syndrome (CAPS) is a condition defined by multiple thromboembolic occurrences within a week period in a patient with positive antiphospholipid antibodies. It has been reported in literature in patients with ulcerative colitis. We will describe an atypical case of ulcerative colitis, with no previous history of antiphospholipid syndrome, who developed CAPS after surgical management of her Leriche syndrome. We also provide a review of the literature to identify the importance of CAPS for clinicians to be aware of this syndrome when the patient is not known to have antiphospholipid syndrome.

Keywords: Catastrophic antiphospholipid syndrome; Antiphospholipid syndrome; Hypercoagulable state; Ulcerative colitis

Abbreviations

Ab2GLP1: Anti-b2 Glycoprotein1; Ab: Antibody; AKI: Acute Kidney Injury; ANA: Antinuclear Antibody; ANCA: Antineutrophil Cytoplasmic Antibody; aPL: Antiphospholipid Antibodies; APS: Antiphospholipid Syndrome; BP: Blood Pressure; CAPS: Catastrophic Antiphospholipid Syndrome; CFA: Common Femoral Artery; CVA: Cerebrovascular Accident; DIC: Disseminated Intravascular Coagulation; DM: diabetes Mellites; GCS: Glasgow Coma Scale; HDL: High Density Lipoprotein; HIT: Heparin Induced Thrombocytopenia; HR: Heart Rate; HTN: Hypertension; IBD: Inflammatory Bowel Disease; ICU: Intensive Care Unit; Ig: Immunoglobulin; IMA: Inferior Mesenteric Artery; LDL: Low Density Lipoprotein; LMWH: Low Molecular Weight Heparin; PCEA: Patient Controlled Epidural Analgesia; POD: Postoperative day; PTT: Partial Thromboplastin Time; SFA: Superficial Femoral Artery; SLE: Systemic Lupus Erythematosus; SMA: Superior Mesenteric Artery; TG: Triglyceride; tPA: tissue Plasminogen Activator; TTE: Transthoracic Echocardiography

Introduction

Failure of multiple organs within a week's period in patients who have antiphospholipid antibodies presents a disastrous situation to surgeons [1]. This phenomenon was named "catastrophic" antiphospholipid syndrome "CAPS" by Asheron, thus the other name "Asheron Syndrome" [1]. The CAPS Registry is an international registry for patients who develop catastrophic APS; it was generated by the European Forum on aPL in the year 2000. Analysis of the registry showed that the predisposing factor was primary APS in 46% of patients, SLE in 40%, lupus-like disease in 5%, and different autoimmune diseases in 9% [2]. It is noteworthy to state that 46% of CAPS patients may present *de novo* without any event of previous thrombosis. However, 53% had precipitating factors [3]. Thankfully, as little as 1% of the patients with anti-phospholipid antibodies present with CAPS [4]. There are many identified precipitating factors for the development of CAPS, these include: infections in 20%, operative procedures in 14%, malignancies in 9%, withdrawal of anticoagulation in 7%, complications of obstetrics in 5%, lupus flares in 4% and the use of oral contraceptive pills in 3% [5,6].

Case Presentation

A 52-year-old female patient presented to our hospital for an elective aortobifemoral bypass for Leriche syndrome. Her past medical history is significant for ulcerative colitis on mesalamine, HTN, DM, and CVA. On postoperative day 2 she developed "Catastrophic Antiphospholipid Syndrome" (CAPS) and passed away 10 days after the surgery.

OPEN ACCESS

Correspondence:
Sarah Assaf, Lebanese American University, Beirut 1102, Beirut, Lebanon,
E-mail: sarah.assaf01@lau.edu

Received Date: *18 Aug 2022*
Accepted Date: *09 Sep 2022*
Published Date: *14 Sep 2022*

Citation:
Assaf S, Daher R, Chemali R, Hayek F. Probable CAPS after Surgical Intervention in a Patient with Known Ulcerative Colitis: A Case Report and Literature Review. Clin Case Rep Int. 2022; 6: 1395.

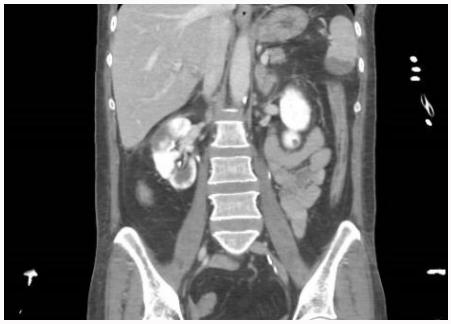

Figure 1: The colon had a tubular lead-pipe appearance on CT scan coronal view.

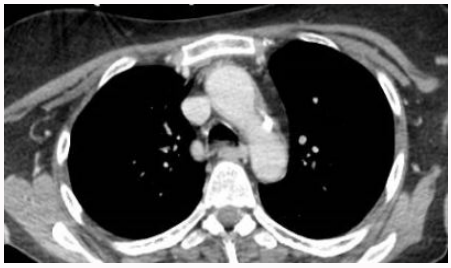

Figure 2: Irregular atheromatous plaque within the aortic arch on CT scan cross section view.

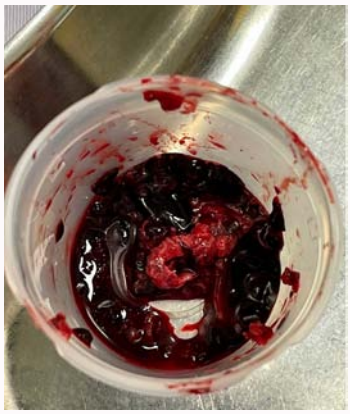

Figure 3: Emboli retrieved via embolectomy using Fogarty catheter after acute aortobifemoral graft occlusion.

Three months prior to presentation, she had presented to the hospital with a thrombotic stroke in her left middle cerebral artery. Thrombolysis with tPA was attempted and failed. Thrombectomy was performed 10 h after the beginning of her symptoms. During thrombectomy through the femoral approach, the wire failed to pass through her common femoral artery bilaterally but was successfully passed through her right brachial artery. Post-thrombectomy she was started on Enoxaparin 40 mg and Aspirin 100 mg daily.

It is worth stating that 11 days prior to her stroke, the patient presented with a 3-week history of diarrhea and was diagnosed with a flare up of her ulcerative colitis and was thus started on Enoxaparin 40 mg daily. Thus, our patient had her stroke despite being on prophylactic anticoagulation.

As the wire failed to pass through her femoral artery (bilaterally) during thrombectomy, CT angiography was performed and demonstrated a complete occlusion of her infrarenal aorta, extending to the common iliac arteries consistent with Leriche syndrome. The inferior mesenteric artery was completely occluded. She had multiple splenic and bilateral renal infarcts. The colon had a tubular lead-pipe appearance (Figure 1). She had an irregular atheromatous plaque within the aortic arch (Figure 2).

Concerning the hypercoagulability workup, the patient had a negative lipid profile, with HDL cholesterol of 32 mg/dl, LDL cholesterol of 24, and TG of 138. Negative ANA, C3, C4, Homocysteine, Protein C, Protein S, Procalcitonin, Antithrombic activity, Factor VIII, p-ANCA and c-ANCA. She also had negative Cardiolipin Ab IgG and IgM, Phospholipid Ab IgG and IgM. Her Lupus anticoagulant was positive, but she was on Enoxaparin, thus it was interpreted as a false positive result, especially with the negative cardiolipin Ab.

She had a Fecal calprotectin of >2100 (high quantity >200 micrograms/g). Ten days after her stroke, she was discharged with right sided hemiplegia and aphasia on Aspirin and Rivaroxaban.

Three months following her CVA, the patient was admitted for an aortobifemoral bypass. Preoperatively, she was switched from Rivaroxabanto enoxaparin. She had Patient Controlled Epidural Anesthesia (PCEA) inserted directly preoperatively for postoperative pain control. Intraoperatively, the right external iliac artery was atherosclerotic. 5000 Units of unfractionated heparin were injected intravenously before clamping of the aorta. An anastomosis was performed between Dacron graft and the aorta, then between the graft and both femoral arteries. At the end of the procedure a positive right and left Common Femoral Artery (CFA) pulse was felt.

Postoperatively, ultrasound showed flow in the SFA and profound a bilaterally. In the ICU, the patient developed altered mental status with worsening of her aphasia. Brain MRI did not show new changes. Enoxaparin was started 40 mg SC BID 5 h post-operatively. Her

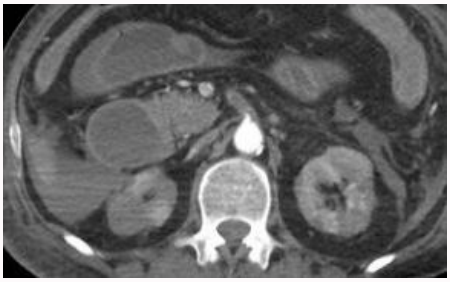

Figure 4: CT angiography of the abdomen showing proximal SMA thrombus.

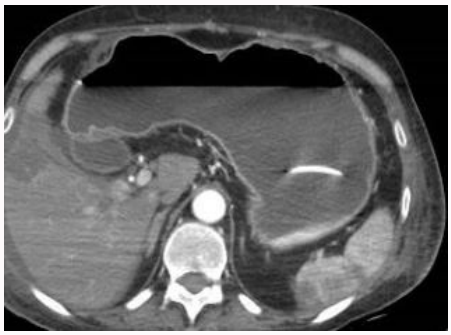

Figure 5: CT angiography of the abdomen showing multiple scattered hepatic infarcts as well as splenic infarcts.

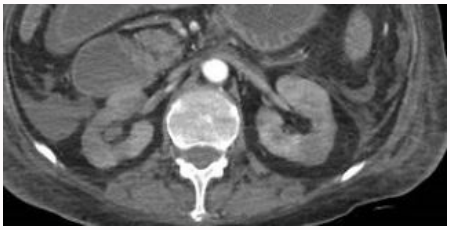

Figure 6: CT angiography showing occlusion of both renal arteries distally with bilateral renal infarcts.

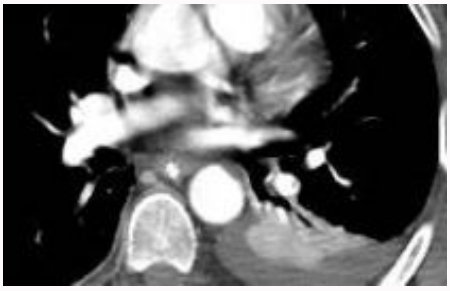

Figure 7: CT angiography showing lower lobar arterial pulmonary emboli.

post-operative course was complicated by acute occlusion of her aortobifemoral bypass on post-operative day 1. Subsequently, a bilateral embolectomy was performed using Fogarty catheter (Figure 3). The patient was then started on unfractionated Heparin IVSP 25000 IU /24 h monitored by PTT level.

Post-embolectomy, the patient developed rhabdomyolysis complicated by AKI, she also had hypotension unresponsive to IV fluids and became tachypneic but was not able to compensate for her metabolic acidosis. She was intubated, started on norepinephrine and antibiotics and on day 3 post-operatively she was started on hemodialysis. Methylprednisolone 100 cc once was given due to a possible component of adrenal insufficiency and was continued at a rate of 50 cc q 6 h. The skin on her right thigh also showed livedo reticularis, but these were not confirmed by biopsy due to financial reasons. She developed thrombocytopenia with a platelet count reaching 49 000/mm^3. Heparin-Induced Thrombocytopenia (HIT) and Disseminated Intravascular Coagulation (DIC) were ruled out as she had a D-dimer of 3.06 and a fibrinogen level of 354. Her mental status was disturbed as she was intubated, off sedation, with a Glasgow Coma Scale (GCS) of 3. Brain CT did not show any changes.

CT angiography showed acute mesenteric ischemia involving the small bowel secondary to a proximal SMA thrombus (Figure 4), multiple scattered hepatic infarcts, occlusion of both renal arteries distally (Figure 5) with increase of the bilateral renal infarcts (Figure 6). Bilateral lower lobar arterial pulmonary emboli (Figure 7). Her Beta 2 Glycoprotein Abs IgM and IgG were positive, Lupus anticoagulant and Anticardiolipin IgG and IgM were negative. She was continued on methylprednisolone 50 mg q 6 h and therapeutic range unfractionated heparin monitored by Anti-factor X a assay. Due to her poor prognosis, her family changed the authorization to comfort care only. She deteriorated and passed away 10 days post-operatively.

Discussion

Asheron et al. proposed criteria for diagnosing CAPS. Certain features were common between these patients: Multiple organs (>2) involved, many small vessels obstruction found histologically and the presence of antiphospholipid antibodies on serology [5] (Table 1).

Table 1: Proposed criteria for the diagnosis of CAPS.

Proposed criteria for the diagnosis of CAPS
1. Vessel occlusions in more than 2 organs found clinically
2. Presence of antiphospholipid antibodies on serology
3. An organ with a small blood vessel occluded on histology
4. Occurrence of the above manifestations in less than a week
5. Definite CAPS involve all four criteria while probable CAPS involve only 3 criteria [5].

These were later validated in the year 2005 by Cervara et al. [6] who found that they are 90.3% sensitive and 99.4% specific [6].

It is of utmost importance to have the diagnosis early; but, due to multiple reasons, it is challenging. Aside from the fact that CAPS is a rare disease, it might not be thought of in patients without a known APS. Even if considered, not all diagnostic criteria may be met; especially in the acute setting [7].

Regarding management of patients with CAPS, early initiation of treatment, within 8 hours of diagnosis, was considered of extreme importance. The first and most important treatment for such patients is therapeutic level anticoagulation [7]. Steroids, anticoagulants and plasma exchange combined provided the highest recovery rate [3].

We present a case of a 52-year-old female, with no previous pregnancy attempts, not known to have APS who was operated on and died after developing probable CAPS. Yet, she is known to have ulcerative colitis. The aim of this case report is to emphasize the importance of identifying CAPS as a differential diagnosis for patients who develop thrombocytopenia with no DIC nor HIT; but, with multiple organ damage and are not known to have APS nor SLE. This is of utmost importance as rapid treatment may save the life of the patient.

We also aim to emphasize the probability of CAPS in the large population of patients with IBD and increase the level of suspicion for CAPS in surgical patients. There is evolving evidence that the pathogenesis of formation of both arterial and venous thrombosis involves antiphospholipid antibodies [9]. In fact, in a study conducted by Kraiem et al. [10], titled antiphospholipid antibodies and procoagulant profile in Tunisians with IBD, the authors concluded that the antiphospholipid auto antibodies, mostly IgA ab2GPI antibodies, were very often found in Tunisian patients with IBD. They suggested more prospective studies be conducted to learn about this marker's exact function and clinical relevance [10].

A case report published by Stammler et al. [11], further supported recommendations that anti- vitamin K medication is the anticoagulant of choice in patients who have all three positive antiphospholipid antibodies [11]. Our patient, who had only 2 of the three antibodies positive, was on Enoxaparin 40 mg daily before she had her stroke. She had her stoke, despite being on enoxaparin which was prescribed to her due to an ulcerative colitis disease flare up. Postoperatively our patient developed CAPS triggered by the surgery.

Importantly, in preparation for her postoperative pain, a PCEA was placed preoperatively. On POD 4, while the patient was on therapeutic anticoagulation, removal of the PCEA was problematic, as it would requires interruption of the anticoagulation due to fear of an epidural hematoma. This is an area of controversy, and further

studies are required for identifying patients who are not fit for epidural pain control due to their hypercoagulable state that might require uninterrupted therapeutic anticoagulation post operatively. Patients with hypercoagulable state could be identified as being a relative contraindication for placement of a preoperative PCEA.

In conclusion, we encountered a case of probable CAPS in a patient not known to have antiphospholipid syndrome but known to have ulcerative colitis. It was a dramatic experience to have a patient develops such a chaotic sequence of events. Despite its rarity, CAPS should be kept in mind as one of the differentials in a patient with known ulcerative colitis who develops thrombocytopenia with multi-organ dysfunction following surgery. The early diagnosis of the disease with early management with uninterrupted therapeutic anticoagulation and steroids could save the life of the patient. Further studies are required for finding the incidence of developing CAPS in patients with inflammatory bowel disease.

Placement of a PCEA is an area of controversy that requires further studying. Surgeons must always keep in mind that surgery acts as a trigger for developing a hypercoagulable state leading to CAPs.

References

1. Asherson RA. The catastrophic antiphospholipid syndrome. J Rheumatol. 1992;19:508-12.
2. Cervera R, Bucciarelli S, Plasín MA, Gómez-Puerta JA, Plaza J, Pons-Estel G, et al. Catastrophic Antiphospholipid Syndrome (CAPS): Descriptive analysis of a series of 280 patients from the "CAPS Registry". J Autoimmun. 2009;32(3-4):240-5.
3. Cervera R. Catastrophic Antiphospholipid Syndrome (CAPS): Update from the 'CAPS Registry'. Lupus. 2010;19(4):412-8.
4. Cervera R, Piette JC, Font J, Khamashta MA, Shoenfeld Y, Camps MT, et al. Antiphospholipid syndrome: Clinical and immunologic manifestations and patterns of disease expression in a cohort of 1,000 patients. Arthritis Rheum. 2002;46(4):1019-27.
5. Asherson RA, Cervera R, Piette JC, Font J, Lie JT, Burcoglu A, et al. Catastrophic antiphospholipid syndrome. Clinical and laboratory features of 50 patients. Medicine (Baltimore). 1998;77(3):195-207.
6. Cervera R, Font J, Gómez-Puerta JA, Espinosa G, Cucho M, Bucciarelli S, et al. Validation of the preliminary criteria for the classification of catastrophic antiphospholipid syndrome. Ann Rheum Dis. 2005;64(8):1205-9.
7. Erkan D, Espinosa G, Cervera R. Catastrophic antiphospholipid syndrome: Updated diagnostic algorithms. Autoimmun Rev. 2010;10(2):74-9.
8. Ruffatti A, De Silvestro G, Marson P, Tonello M, Calligaro A, Favaro M, et al. Catastrophic antiphospholipid syndrome: Lessons from 14 cases successfully treated in a single center. A narrative report. J Autoimmun. 2018;93:124-30.
9. Salmon JE, de Groot PG. Pathogenic role of antiphospholipid antibodies. Lupus. 2008;17(5):405-11.
10. Kraiem I, Hadhri S, Ben Rejeb M, Ifa L, Jmaa A, Ajmi S, et al. Antiphospholipid antibodies and procoagulant profile in Tunisians with inflammatory bowel diseases. Clin Appl Thromb Hemost. 2016;22(8):734-42.
11. Stammler R, Legendre P, Cacoub P, Blanche P, Piette JC, Costedoat-Chalumeau N. Catastrophic antiphospholipid syndrome following the introduction of rivaroxaban. Lupus. 2020;29(7):787-90.