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SERONEGATIVE CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME IN A YOUNG FEMALE PRESENTING WITH A HEADACHE

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☐ **Abstract—Background: Catastrophic antiphospholipid syndrome (CAPS) is a rare disease that causes rapid vascular occlusion in multiple organ systems. Initial presentation varies depending on the organs affected. Although headache is a common complaint in the emergency department (ED), it is a very rare presentation of CAPS. Case Report: A 43-year-old previously healthy woman presented to the ED with severe headache. Subarachnoid hemorrhage was excluded and she was discharged home. She returned 36 h later with diabetic ketoacidosis, hyperthyroidism, and thrombosis in her cerebral venous sinus, aorta and splenic artery. She was treated with heparin, steroids, plasmapharesis, and i.v. immunoglobulin, after which she improved. This constellation of symptoms is highly suggestive of CAPS initiated by a polyglandular autoimmune syndrome, despite negative serology. Why Should an Emergency Physician Be Aware of This?: Although a rare cause of headache, CAPS is a potentially fatal disease that requires early identification and initiation of appropriate treatment. © 2015 Elsevier Inc.**

☐ **Keywords—catastrophic antiphospholipid syndrome; antiphospholipid syndrome; emergency services; autoimmune polyglandular syndrome**

INTRODUCTION

One unusual cause of headache with significant clinical consequences is catastrophic antiphospholipid syndrome (CAPS). Antiphospholipid antibody syndrome (APS) is an autoimmune disease characterized by antibodies against the antiphospholipid complex, which presents as thrombosis. CAPS is a very rare sequela, occurring in < 1% of patients with APS, in which vascular occlusion occurs in three or more locations simultaneously (Table 1) (1–3). CAPS has a high morbidity and mortality, with death occurring in up to 44% of patients (4). Initial presentations vary but rarely occur as cerebral venous thrombosis, as in our case. Clinical identification and early treatment by emergency medicine physicians remains essential.

CASE REPORT

A 43-year-old woman from Afghanistan with recent onset of vitiligo but no other known medical history first presented to the emergency department (ED) with the "worst headache of her life." A noncontrast computed tomography (CT) scan of her head and lumbar puncture (LP) were performed to rule out subarachnoid hemorrhage. Both were unremarkable. The complete blood count and basic metabolic panel were within normal limits except for a serum glucose of 337 mg/dL. Her headache improved with i.v. fluids and morphine and she was discharged home. She returned to the ED 36 h later with altered mental status and vomiting.

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Table 1. International Consensus on Preliminary Criteria for the Classification of the Catastrophic Antiphospholipid Syndrome

Criteria
1. Evidence of thrombosis in three or more organs, systems, or tissues
2. Development of manifestations simultaneously or in < 1 week
3. Confirmation by histopathology of small vessel occlusion in at least one organ or tissue
4. Laboratory confirmation of the presence of antiphospholipid antibodies (lupus anticoagulant or anticardiolipin antibodies)

Classification
- Definite CAPS
  - Requires all four criteria
- Probable CAPS
  - a. All four criteria, except for only two organs, systems, or sites of tissue involvement **or**
  - b. All four criteria, except for the laboratory confirmation at least 6 weeks apart due to the early death of the patient never tested for antiphospholipid syndrome before CAPS or
  - c. Criteria 1, 2, and 4 above or
  - d. 1, 3, and 4 and the development of a third event in > 1 week but < 1 month, despite anticoagulation

CAPS = catastrophic antiphospholipid syndrome.

On examination, her rectal temperature was 35.6°C, heart rate was 124 beats/min, blood pressure was 134/93 mm Hg, respiratory rate was 40 breaths/min, and oxygen saturation was 100% on room air. She was extremely agitated and not following commands. Her examination was notable for diaphoresis, an irregularly irregular cardiac rhythm with a rapid rate, thyromegaly, tenderness to palpation in the left upper quadrant of her abdomen, and areas of hypopigmentation on her legs, arms, and face.

An electrocardiogram was remarkable for atrial fibrillation with rapid ventricular response. Laboratory tests were significant for a white blood count of $21.8 \times 10^3/\mu L$, serum glucose of 560 mg/dL, creatinine of 1.9 mg/dL, bicarbonate of < 5 mmol/L, and an anion gap acidosis > 30 with a normal lactate. Her thyroid-stimulating hormone was < 0.01 mIU/L. She was intubated for airway protection. An abdominal CT with contrast revealed a 1-cm thrombus in the abdominal aorta and a splenic artery thrombus with splenic infarction (Figure 1A and 1B). A repeat noncontrast head CT remained unremarkable. Contrast-enhanced magnetic resonance imaging (MRI) and magnetic resonance venography revealed thrombi in the left transverse and sigmoid sinus (Figure 2). A repeat LP was performed, which showed elevations in opening pressure, protein level, and red blood cells without xanthochromia, consistent with venous thrombosis.

The patient was started on a heparin infusion for multiple thrombi, an insulin infusion for diabetic ketoacidosis (DKA), a β-blocker for atrial fibrillation, high-dose steroids for possible cerebritis, methimazole for hyperthyroidism, and broad-spectrum antibiotics for possible occult infection after blood, urine, and cerebrospinal fluid cultures were obtained. She was admitted by the internal medicine service to the intensive care unit (ICU) for further management.

During her course in the ICU, she remained unresponsive to verbal or nonpainful tactile stimuli despite resolution of her DKA, treatment for hyperthyryroidism, anticoagulation, and high-dose steroids. Her cultures were negative for bacterial growth. Endocrinology,

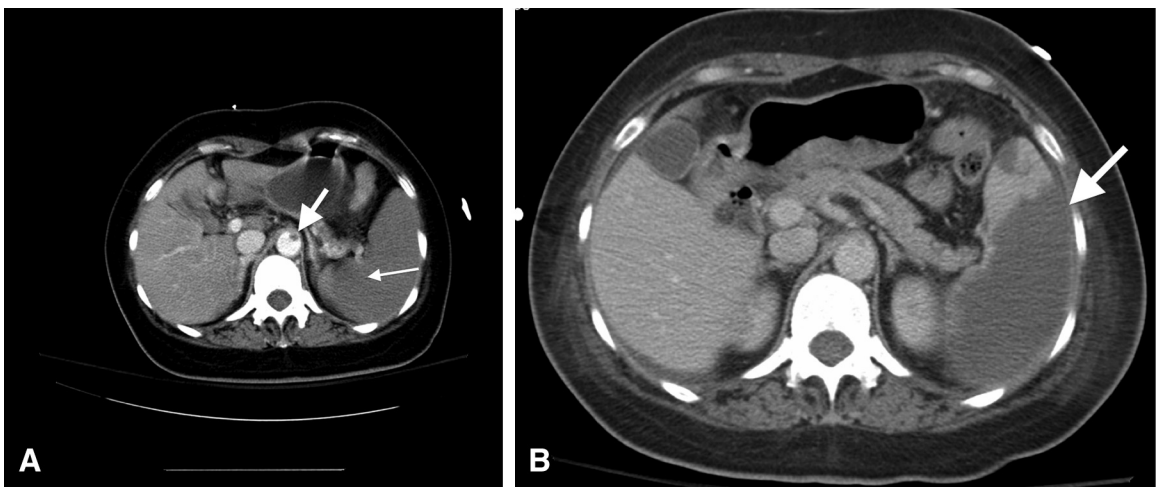

Figure 1. (A) Computed tomography scan of the abdomen with i.v. contrast performed on hospital day 1 showing 1-cm filling defect in the anterior aspect of the abdominal aorta (thick arrow) and hypodense spleen representing splenic infarction (thin arrow). Poor visualization of splenic artery. (B) Computed tomography scan of the abdomen with i.v. contrast completed on hospital day 3 showing extension of splenic infarct.