

A CATASTROPHIC SERIES OF EVENTS

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INTRODUCTION

Catastrophic Anti-phospholipid Syndrome (CAPS) is a rare life-threatening condition that occurs in <1% of patients with antiphospholipid syndrome, and carries a mortality rate greater than 50%. The condition involves widespread thrombotic disease with multi-organ failure.

CASE PRESENTATION

We present a 62yo female with a prior history of anti-cardiolipin antibody who initially presented to the ED for evaluation of back pain, was discharged with a presumptive diagnosis of musculoskeletal pain, and later admitted the next day after a fall. On presentation she had leukocytosis-WBC16.5, severe thrombocytopenia with PLT count of 21, rising troponin levels-3.8 as well as LFT elevations AST576, ALT280, Alkaline Phos 166, and Acute Kidney Injury with Cr of 2.45. She was transferred to the ICU. On hospital day 2 the patient developed L sided weakness and

MRI of the brain revealed subacute ischemia in the right parietal-peritrial white matter. The patient was treated, per current recommendations, with high dose steroids (methylprednisone1000mg for 3 days) and plasma exchange. Symptoms and labs continued to improve during admission. She was started on Rituximab, as this has been shown to rapidly improve severe thrombocytopenia associated with CAPS. She recovered with some focal deficits and was discharged to inpatient rehab.

PATIENT UPDATE:
SHE EVENTUALLY IMPROVED WITH INPATIENT REHAB BUT NEVER BACK TO HER BASELINE. AFTER SEVERAL MONTHS SHE PRESENTED TO THE HOSPITAL ONCE AGAIN WITH SYMPTOMS OF CAPS AND WAS TRANSITIONED TO COMFORT CARE AND EVENTUALLY EXPIRED.

SUMMARY

CAPS is a rare condition, and a high level of suspicion is needed for diagnosis. Current literature suggests that the development of CAPS is caused by a combination of a gene mutation and an environmental trigger, often an infection, in patients with antiphospholipid- syndrome. Literature also suggests that a genetic predisposition may account for why so few people with antiphospholipid syndrome develop CAPS. Our patient was interesting as we were unable to determine an actual precipitating factor that led to CAPS. Patient fulfilled criteria for definite CAPS with symptoms presenting within 1 week, involvement of at least 3 organ systems, positive anti-cardiolipin antibody and histo-pathologic evidence of small vessel occlusion.

DIAGNOSTIC CRITERIA FOR CAPS

1. Evidence of involvement of 3 or more organ systems
2. Development of manifestations simultaneously or within a week of each other
3. Confirmation by histopathology of small vessel occlusion in at least one organ or tissue
4. Laboratory confirmation of the presence of aPL (antiphospholipid antibody)

Adapted from: Asherson, RA, Cervera, R, de Groot, PG, et al. Lupus 2003; 12:530.

RESOURCES

Catastrophic Antiphospholipid Syndrome
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