Tumid lupus erythematosus–like pseudovasculitic lesions in catastrophic antiphospholipid syndrome

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INTRODUCTION

Catastrophic antiphospholipid syndrome (CAPS) is a rare but life-threatening disease entity characterized by widespread vascular thromboses causing multiorgan failure in the setting of antiphospholipid antibody positivity.[1] CAPS is the most severe subset of antiphospholipid syndrome (APS), an autoimmune clotting disorder in which the body attacks its own phospholipids. CAPS can present with a variety of pseudovasculitic skin findings. We report on a CAPS patient with tumid lupus erythematosus (LE)-like lesions.

Abbreviations used:

APS:	antiphospholipid syndrome
CAPS:	catastrophic antiphospholipid syndrome
LE:	lupus erythematosus

CASE REPORT

A woman in her 40s was admitted from the emergency department for acute right epigastric pain with lesions on computed tomography of the liver concerning for possible abscesses. The

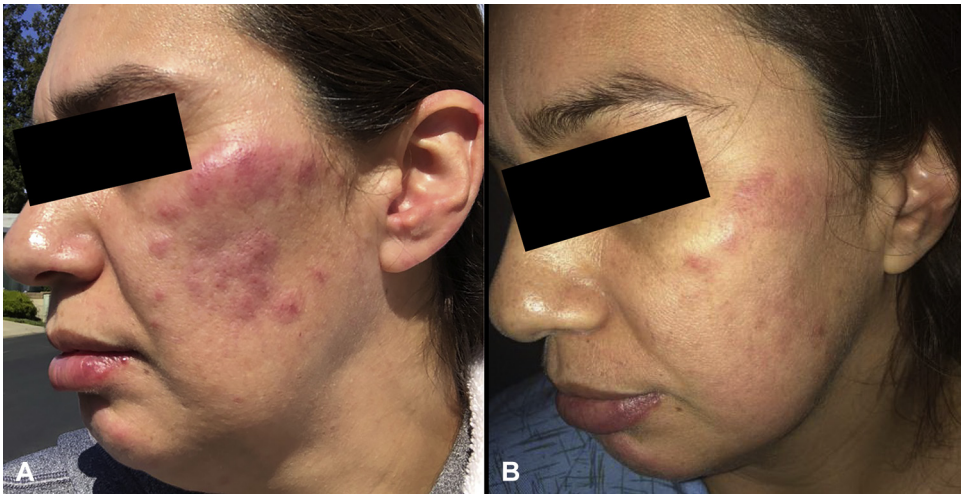

Fig 1. **A**, Dusky-red edematous papules and plaques on the right cheek 3 days before hospital admission. Similar lesions appeared on the left cheek and upper chest (not shown). **B**, Lesions on day of admission.

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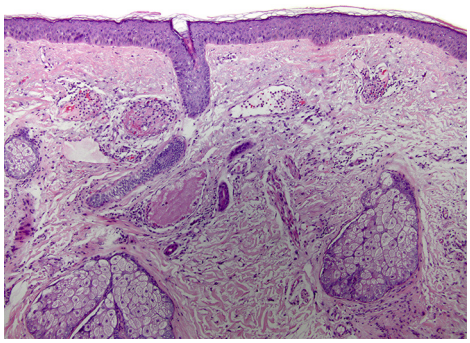

Fig 2. Fibrin thrombi and sparse neutrophils in the upper dermis. (Hematoxylin-eosin stain; original magnification: ×40.)

dermatology department was consulted for evaluation of an asymptomatic eruption on her face and chest. It started on both helices 1 month prior, then crusted and healed. Three days before admission, she experienced abdominal pain with red raised lesions on her face and chest. The lesions were not itchy, tender, or scaly. She started oral contraceptives 1 month prior. She had an 11-week miscarriage 3 years prior and a remote history of childhood scleroderma but was otherwise healthy. On examination, she was noted to have dusky-red succulent papules and plaques scattered on her cheeks and upper chest reminiscent of tumid LE (Fig 1). Antinuclear antibody, anti-dsDNA, and anti-SSA were positive, but anti−Scl-70 and anticentromere were negative. Skin biopsy of the left malar eminence was obtained.

The following day, the biopsy found fibrin thrombi and sparse neutrophils in the upper dermis, consistent with a coagulopathy rather than tumid LE (Fig 2). Coagulopathy was suspected and relevant laboratory studies were ordered.

On hospital day 3, she had substernal chest pain and was transferred to the medical intensive care unit for sepsis and possible myocardial infarction (elevated troponin 2682). Her laboratory values showed positive antibodies (anticardiolipin, anti−β-2 glycoprotein, lupus anticoagulant) and negative antineutrophil cytoplasmic antibodies. Computed tomography of the abdomen showed bilateral adrenal hemorrhages and multifocal liver thromboses. CAPS diagnosis was made given multiorgan involvement (cardiac, adrenal, hepatic, cutaneous), rapid progression (<1 week), and positive antibodies (anticardiolipin, anti−β-2 glycoprotein, lupus anticoagulant) according to the 2003 published international consensus on CAPS diagnosis (Table I).[2] Anticoagulation, intravenous methylprednisolone, plasmapheresis, mycophenolate mofetil, and hydroxychloroquine were initiated. Upon discharge, she was clinically improved with no clinical evidence of ongoing thrombosis.

Table I. Preliminary criteria for the classification of catastrophic APS

Definite diagnosis of catastrophic APS

1. Evidence of involvement of 3 or more organs, systems, and/or tissues*
2. Development of manifestations simultaneously or in less than a week
3. Confirmation by histopathology of small vessel occlusion in at least 1 organ or tissue†
4. Laboratory confirmation of the presence of antiphospholipid antibodies (lupus anticoagulant and/or anticardiolipin antibodies)‡

Probable diagnosis of catastrophic APS

- All 4 criteria, except for only 2 organs, systems and/or tissues involvement
- All 4 criteria, except for the absence of laboratory confirmation at least 6 weeks apart due to the early death of a patient never tested for antiphospholipid antibodies before the catastrophic APS
- 1, 2, and 4
- 1, 3, and 4 and the development of a third event in more than a week but less than a month, despite anticoagulation

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*Usually, clinical evidence of vessel occlusions, confirmed by imaging techniques when appropriate. Renal involvement is defined by a 50% increase in serum creatinine, severe systemic hypertension (>180/100 mm Hg), and/or proteinuria (>500 mg/24h).
†For histopathologic confirmation, significant evidence of thrombosis must be present, although vasculitis may coexist occasionally.
‡If APS was not previously diagnosed in the patient, the laboratory confirmation requires that presence of antiphospholipid antibodies must be detected on 2 or more occasions at least 6 weeks apart (not necessarily at the time of the event), according to the proposed preliminary criteria for the classification of definite APS.

DISCUSSION

Skin involvement occurs in up to 49% and 70% of APS and CAPS patients, respectively,[3] and has been characterized as pseudovasculitis, defined as lesions that evoke clinical consideration of vasculitis but for which vasculitis is histologically excluded.[4] Pseudovasculitic skin findings are also found in occlusive vasculopathies (ie, livedo reticularis, livedo racemosa, livedoid vasculopathy, retiform purpura, digital ulcerations, necrosis, thrombophlebitis) or noninflammatory hemorrhagic disorders (ie, petechiae, purpura/ecchymosis, splinter hemorrhages).[4] Anetoderma-like lesions, discoid lupus erythematosus, Degos-like lesions, and painful

papules and nodules have also been reported in APS[5] but are rarely found in other occlusive vasculopathies. Tumid LE-like or urticarial papules and plaques were not described in the recently published report from the International CAPS Registry.[6] Our case documents tumid LE-like skin findings within the pseudovasculitic spectrum of CAPS, and possibly APS more generally.

This case highlights the important role dermatologists have in the complicated diagnosis of CAPS. High index of suspicion and early diagnosis by the dermatology consult team were likely critical for this patient's survival. Therefore, it is crucial that we continue to catalogue the various skin manifestations of CAPS to improve the diagnostic assessment of this life-threatening disease. We believe tumid LE-like lesions should be classified as APS pseudovasculitic lesions.

REFERENCES

1. Nayer A, Ortega LM. Catastrophic antiphospholipid syndrome: a clinical review. *J Nephropathol*. 2014;3(1):9-17.
2. Asherson RA, Cervera R, de Groot PG, et al. Catastrophic antiphospholipid syndrome: international consensus statement on classification criteria and treatment guidelines. *Lupus*. 2003;12(7):530-534.
3. Asherson RA, Frances C, Iaccarino L, et al. The antiphospholipid antibody syndrome: diagnosis, skin manifestations and current therapy. *Clin Exp Rheumatol*. 2006;24(1 Suppl 40): S46-S51.
4. Carlson JA, Chen KR. Cutaneous pseudovasculitis. *Am J Dermatopathol*. 2007;29(1):44-55.
5. Gibson GE, Su WP, Pittelkow MR. Antiphospholipid syndrome and the skin. *J Am Acad Dermatol*. 1997;36(6 Pt 1): 970-982.
6. Rodriguez-Pinto I, Moitinho M, Santacreu I, et al. Catastrophic antiphospholipid syndrome (CAPS): descriptive analysis of 500 patients from the International CAPS Registry. *Autoimmun Rev*. 2016;15(12):1120-1124.