

CASES FOR DIAGNOSIS

Violaceous Papules and Nodules on the Palmar Aspects of the Hands

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Patient History

The patient was a 31-year-old woman with a history of abortion in the 16th week of pregnancy and diagnosed with possible antiphospholipid syndrome (APS) during her second pregnancy, based on the incidental finding of positive antiphospholipid antibodies and her previous history. In the third month of pregnancy, she initiated antiplatelet therapy with acetylsalicylic acid (100 mg/d). In the 35th week, she was admitted for scheduled caesarean section due to hypertension and intrauterine growth retardation of the fetus. Swelling and painful cutaneous lesions in both hands were observed 2 days after delivery.

Physical Examination

The physical examination revealed edema on the palms and anterior aspects of the fingers, with erythematous violaceous plaques and infiltrated areas that were sensitive to palpation and showed a purplish hue (Figure 1). There was no bleeding under the nails or cutaneous lesions in any other area. No significant finding except for blood pressure of 160/95 mm Hg was seen in the general physical examination.

Additional Examinations

Complete blood count: hemoglobin, 11.5 g/dL; leukocytes, 10 400 cells/ μ L (normal leukocyte differential); platelets, 75 000 cells/ μ L; erythrocyte sedimentation rate, 109 mm/h. *Biochemistry:* normal, including liver and renal profile. *Coagulation:* fibrinogen, 536 mg/dL (normal range, 200-400 mg/dL); activated partial thromboplastin time, 47.9 s (normal range, 26-34 s); activated partial thromboplastin ratio, 1.55 (normal range, 0.8-1.3); prothrombin time and international normalized ratio normal. *Urine sediment:* proteins +++, all other findings normal. *Immunology:* negative antinuclear antibodies and rheumatoid factor; normal values of C3 and C4 complement; positive lupus anticoagulant at high titer; anticardiolipin antibodies, IgG >200 U/mL and IgM 47 U/mL.

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Figure 1.



Figure 2.
Hematoxylin-eosin,
 $\times 10$. Insert, $\times 40$.

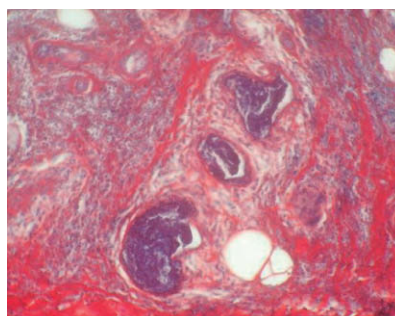


Figure 3.
Phosphotungstic
acid-hematoxylin
staining, in which
the fibrin thrombi are
stained blue ($\times 40$).

Anatomical Pathology

Acral skin with noninflammatory thrombotic vascular disease was observed. The blood vessels of the middle dermis were completely occluded (Figures 2 and 3). The direct immunofluorescence study was negative.

What is your diagnosis?

Diagnosis

Primary APS with cutaneous lesions and preeclampsia

Course

The cutaneous lesions resolved spontaneously within a few days, with the elevated blood pressure figures and the proteinuria disappearing within a few weeks and thrombocytopenia also improving. The patient is currently asymptomatic and receiving antiplatelet therapy.

Comment

APS is a multisystem disease characterized by the presence of prothrombotic autoantibodies against cell membrane phospholipids.¹ It may be primary (more common) or secondary to various diseases, the most significant of which is systemic lupus erythematosus (SLE).¹ Among patients who present with primary APS, very few will later develop manifestations of lupus,² although long-term follow-up is recommended (a 5-year period has been suggested, but there is no consensus).³

There are a number of clinical manifestations that depend on the site of the thrombosis.^{1,4} The most common are arterial or venous thrombosis of the limbs, followed by central nervous system thrombosis and intrauterine deaths due to placental and fetal thrombosis. During pregnancy, APS can also cause fetal distress, intrauterine growth retardation, prematurity, and pregnancy toxemias.

The cutaneous lesions of APS are extremely varied and include livedo reticularis, ulcers, necrosis, thrombophlebitis, splinter hemorrhages,^{1,4,5} and, as in our patient, painful purpuric papulonodular lesions.⁶ Pathology findings are usually typical of noninflammatory obstructive vascular disease.

According to classic diagnostic criteria, APS is diagnosed when at least one of the following is present: (1) arterial or venous thrombosis, (2) repeated abortions and/or fetal deaths (at least 2), and (3) thrombocytopenia, along with (4) detection of antiphospholipid antibodies on at least 2 occasions in more than 3 months. These criteria were reviewed in Japan in 1998, proposing that the clinical evidence should be the first 2 only and that the presence of only 1 antiphospholipid antibody on 2 occasions 6 weeks apart would be sufficient.⁷

Treatment mainly consists of preventing new thrombosis, with long-term anticoagulation indicated. In obstetric APS (in which the clinical manifestations occur only in pregnancy), the best approach is still under discussion, although most authors recommend anticoagulation only during pregnancies, maintaining only antiplatelet agents outside the pregnancies.

Differential Diagnosis

APS Secondary to SLE

This was the main differential diagnosis in our case; until the results were obtained, the clinical symptoms could have been considered due to SLE exacerbation with nephropathy, intrauterine growth retardation, anemia, and thrombocytopenia. SLE can cause small-vessel vasculitis and, in this context, may produce lesions such as those seen in our patient.

Disseminated Intravascular Coagulation

Disseminated intravascular coagulation is a severe, generalized coagulation disorder that can be triggered by various obstetric problems. The cutaneous symptoms consist of hemorrhagic plaques with irregular borders and symmetric distribution on the face, limbs, breasts, pressure areas, etc. Biopsy reveals vascular occlusion by fibrin thrombi, with little inflammation.

Other Noninflammatory Obstructive Vascular Diseases

The differential diagnosis should be done histologically and should include white atrophy, thrombotic thrombocytopenic purpura, Sneddon syndrome, essential thrombocytosis, cutaneous necrosis caused by warfarin, cryoglobulinemia, bacterial endocarditis, C- or S-protein deficiency, and heparin therapy.

Conflicts of Interest

The authors declare no conflicts of interest.

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