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,\(^7\) 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 antithrombophilic 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 antithrombophilic 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.

References