

## CASES FOR DIAGNOSIS

# Erythematous Violaceous Nodules and Telangiectasis on the Thighs

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## Patient History

An 85-year-old woman with a history of diabetes mellitus and ischemic heart disease consulted for low-grade fever, asthenia, and painful cutaneous lesions on the thighs that had begun 7 months previously.

## Physical Examination

The lesions consisted of papules, plaques, and indurated, erythematous violaceous nodules, with a linear and reticular distribution (Figure 1). The left thigh presented a hot, erythematous plaque filled with numerous telangiectases on the surface (Figure 2). No enlarged lymph nodes were palpated in accessible regions, and no pathological findings were observed in the abdominal examination. The neurological examination was normal.

## Additional Examinations

The complete blood count and blood biochemistry were normal except for an increased erythrocyte sedimentation rate (39 at 1 hour) and elevated lactate dehydrogenase (695 IU/L). The chest x-ray was normal. Cranial computed tomography (CT) identified focal hypodensity in the right lenticular nucleus consistent with a lacunar infarction. Chest and abdominal CT scans showed a nodule of 3 × 2 cm in the left adrenal gland that probably was an adrenal adenoma.

## Histopathology

The skin biopsy showed dilated vessels in the mid-dermis and deep dermis with prominent endothelia and occupied



Figure 1.



Figure 2.

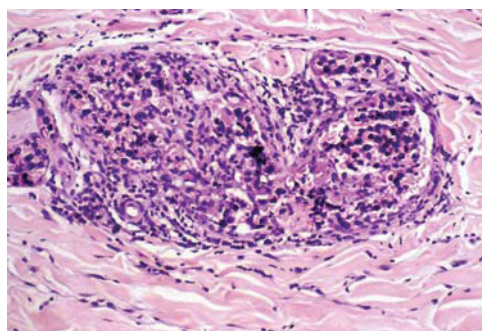


Figure 3.  
Hematoxylin-eosin, ×20.

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by a noncohesive mass of atypical cells with large, pleomorphic, hyperchromatic nuclei, and abundant mitotic figures (Figure 3).

What is your diagnosis?

## Diagnosis

Intravascular large B-cell lymphoma

## Course and Treatment

The immunohistochemical study revealed that the atypical cells confined to the interior of the vessels were intensely positive for CD20. T-cell markers, myeloperoxidase, anion exchangers AE1/AE2, CD34, and the serum protein S-100 were all negative. Bone marrow biopsy was normal. The patient was treated with oral corticosteroids. Four months after the diagnosis, she presented asthenia, double vision, cognitive deterioration, and worsening of the cutaneous lesions, and died 1 month later.

## Comment

Intravascular large B-cell lymphoma is a rare subtype of extranodal diffuse large B-cell lymphoma that is characterized by the location of neoplastic cells, in the lumen of capillaries and postcapillary venules. It has usually spread extensively by the time of diagnosis. The symptoms result from vascular occlusion and can be highly heterogeneous. More than half the cases are diagnosed post-mortem and most patients present systemic symptoms, mainly fever.<sup>1</sup> Between 34% and 85% of cases present neurological symptoms (focal deficits, aphasia, dysarthria, apraxia, confusion symptoms, dementia, epilepsy, myelopathy, visual disturbances, vertigo).<sup>1,2</sup> Cutaneous lesions are the initial sign in 39% of patients and can be single or multiple; they are usually found on the thighs, legs, arms, abdomen, or breasts. Violaceous plaques, orange peel-like edema, painful bluish-red nodules, ulcerated nodules, erythematous desquamative plaques, and telangiectases have been described.<sup>1</sup>

A third of all cases involve the liver, spleen, and bone marrow and exhibit pancytopenia. Lymph node involvement is rare (11%).<sup>1</sup> Other less common clinical manifestations are dyspnea, pulmonary hypertension,<sup>1</sup> syndrome of inappropriate antidiuretic hormone secretion, hypopituitarism, adrenal insufficiency, hyperprolactinemia, increased corticotropin,<sup>3</sup> cytopenias, hypoalbuminemia, and monoclonal gammopathy.<sup>1</sup>

An "Asian variant" has been reported among Japanese individuals; this condition manifests with hemophagocytic

syndrome, bone marrow invasion, enlarged liver and spleen, anemia, thrombocytopenia, and fever.<sup>4</sup>

The histopathological study shows large tumor cells with vesicular, pleomorphic nuclei, prominent nucleolus, and numerous mitotic figures occupying the lumen of small vessels.<sup>1</sup> Tumor cells have been described inside cutaneous or hepatic hemangiomas, renal cell carcinoma, and prostatic hyperplasia.<sup>1</sup> It is usually associated with B-cell phenotype; T-cell phenotype is rare. Genotyping usually reveals monoclonal rearrangement of the gene for immunoglobulin heavy chains. Gene rearrangement of the T-cell receptor has been reported less frequently.<sup>1</sup> Karyotyping shows structural abnormalities in chromosomes 1, 6, and 18.<sup>5</sup>

This is an aggressive process in which the treatment of choice is anthracycline-based chemotherapy. If the central nervous system is affected, methotrexate or cytarabine should also be used. Radiation therapy is reserved for elderly patients with a single cutaneous lesion or as consolidation treatment after chemotherapy in the case of central nervous system involvement or extranodal disease.<sup>6</sup>

## Conflicts of Interest

The authors declare no conflicts of interest.

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