

Behçet's Disease and Periarteritis Nodosa with Cutaneous Lesions

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Figure 1. Erythematous nodules on the anterior side of the leg.

To the Editor:

Behçet disease is characterized by a triad of symptoms—mouth and genital sores, cutaneous lesions, and ocular involvement. Joint, neurological, vascular, gastrointestinal, or pulmonary involvement may also be observed.¹⁻³

A 38-year-old woman consulted for lesions on the legs that had appeared 10 days earlier. These were various poorly circumscribed erythematous nodules, hot to touch and painful on palpation (Figure 1). The patient had developed a fever of up to 38.5°C from 5 days before the consultation.

A biopsy showed a thrombosed medium-size vessel below intact epidermis and dermis. The wall of this vessel presented necrosis and severe inflammatory infiltrate consisting mainly of neutrophils, but also of lymphocytes and eosinophils. The inflammatory infiltrate extended to the lobules of the subcutaneous cell tissue, in the form of panniculitis of lobular predominance (Figure 2). Orcein staining showed that this was an arteriole. These histological findings were consistent with periarteritis nodosa.

The patient returned earlier than scheduled, because she had developed

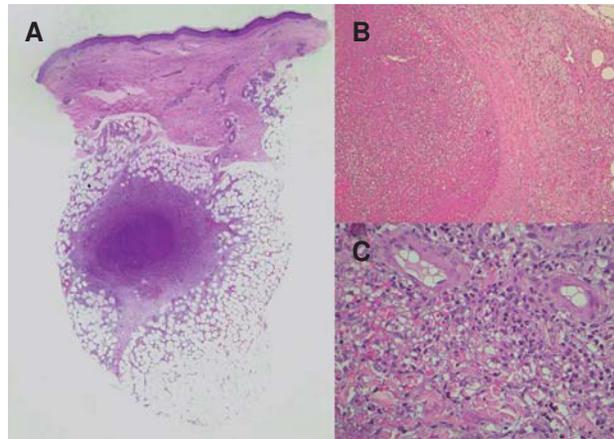


Figure 2.

A. Thrombosed medium-sized vessel in the subcutaneous cell tissue. Hematoxylin-eosin, panoramic view. B. Necrosis in vessel wall with inflammatory infiltrate. Hematoxylin-eosin, $\times 20$. C. Abundant neutrophils in the inflammatory infiltrate. Hematoxylin-eosin, $\times 40$.

oral and genital lesions a few days earlier. There were various painful ulcers on the hard palate, with a yellowish membrane on the surface and an erythematous base (Figure 3). The vulva presented a 1-cm lesion of the same characteristics. The patient reported that she had a 3-year history of recurrent outbreaks of these types of painful lesions in the oral and genital mucosa.

Based on established diagnostic criteria, the patient was diagnosed with Behçet disease with cutaneous periarteritis nodosa lesions. For 1 year, the patient had experienced repeated outbreaks of erythema nodosum, cutaneous papulopustular lesions, and mouth and genital sores, requiring prolonged courses of oral prednisone to control the lesions and colchicine as maintenance therapy.

Erythema nodosum-like lesions are a common cutaneous manifestation in Behçet disease. They are clinically characterized by hot, painful erythematous nodules located mainly on the anterior side of the legs. From the histopathological point of view,

these lesions are not entirely characterized. However, various articles indicate that this type of lesion, clinically indistinguishable from a typical erythema nodosum, presents a different histopathology from this kind of panniculitis. Common findings are inflammatory infiltrate in the dermis, panniculitis of lobular predominance (but also mixed), and vasculitis, both leukocytoclastic and lymphocytic.^{4,5}

Periarteritis nodosa affects small and medium-size arteries and its characteristic histopathology consists of necrotizing vasculitis with predominantly neutrophilic infiltrate



Figure 3. Mouth sores on the palate.

and focal lobular panniculitis near affected vessels. Only a few cases of Behçet disease with erythema nodosum-like lesions where the histopathology showed typical findings of periarthritis nodosa have been described.^{6,7} In our case, we describe a patient who developed erythema nodosum-like cutaneous lesions with histological characteristics of periarthritis nodosa at the time she was diagnosed with Behçet disease. Periarthritis nodosa lesions appear to be a marker of the severity of Behçet disease.⁷ We believe that the onset of this type of lesions in our patient implied worsening of the course of the disease, which had gone almost unnoticed until that time.

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