JAMA Dermatology Clinicopathological Challenge

Cutaneous Nodules and Erythematous Plaques on the Extremities

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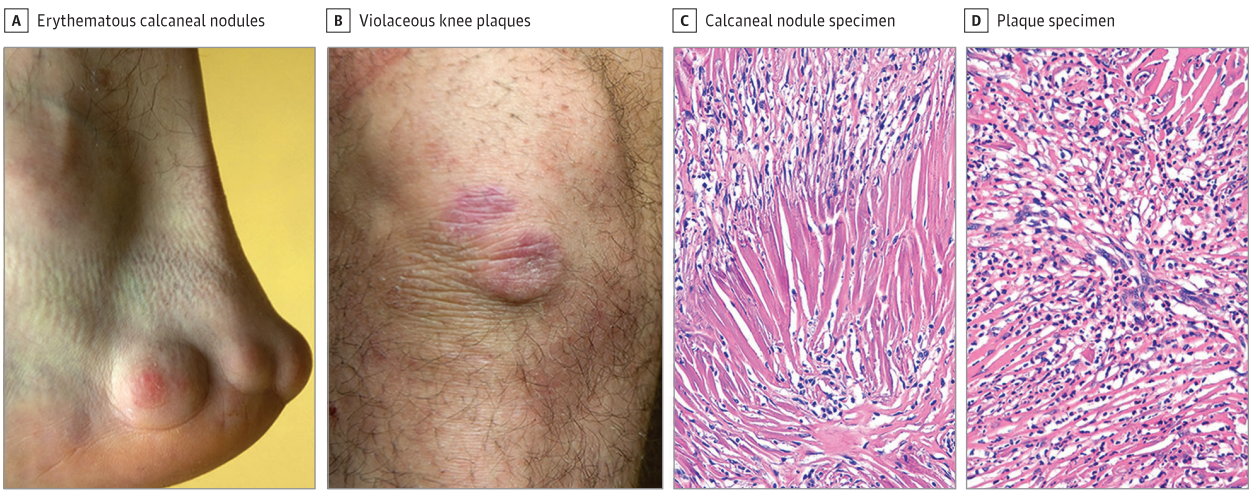

Figure. A, Multiple asymptomatic erythematous nodules measuring 1.5 to 3.0 cm at presentation. B, Multiple violaceous plaques measuring 2 to 3 cm on the extensor surface of the knees. C, Excisional biopsy specimen of a calcaneal nodule (hematoxylin-eosin, original magnification ×20). D, Excisional biopsy specimen of a plaque (hematoxylin-eosin, original magnification ×20).

A healthy man in his 20s presented with a several-month history of asymptomatic, slightly erythematous cutaneous nodules, measuring 1.5 to 3.0 cm, over the calcanea (**Figure**, A). Physical examination revealed violaceous plaques, measuring 2 to 3 cm, on the extensor surface of the knees, bilaterally (Figure, B). Excisional biopsy specimens were obtained (Figure, C and D).

WHAT IS YOUR DIAGNOSIS?

A. Elastolytic granuloma

B. Knee-located erythema elevatum diutinum (EED) with a calcaneal late-stage nodular component

C. Epithelioid sarcoma

D. Storiform collagenoma

Diagnosis

B. Knee-located EED with a calcaneal late-stage nodular component

Microscopic Findings

Histopathological examination of the genicular plaques revealed a dense neutrophilic infiltrate within and around small vessels with micro-abscesses in the papillary and reticular dermis; deposits of fibrin and leukocytoclasia were also present. For the calcaneal nodules, in contrast, a central hypocellular dermal proliferation of thickened and hyalinized collagen bundles with a distinctive storiform pattern was surrounded by multiple foci of concentric "onion-skin" fibrosis with interspersed neutrophils around small vessels.

Discussion

A rare form of cutaneous vasculitis, EED usually affects adults. According to the stage of the lesions, the clinical and histologic features of the disease vary. Early-stage EED is characterized by symmetrical and persistent papules and plaques located on the extensor surfaces of the extremities; histologic sections commonly reveal vascular infiltration in the upper and mid dermis with predominant neutrophils and lesser numbers of lymphocytes, eosinophils, and plasma cells; leukocytoclasia is commonly observed. Such lesions may be histologically indistinguishable from primary vasculitides. However, the typical clinical localization and the absence of systemic involvement are clue features of EED. Some scholars consider EED and granuloma faciale on a spectrum; however, the different localization and the predominance of neutrophils rather than eosinophils in the present case point to the diagnosis of EED.

Late-stage EED has been documented in few reports[1-7] and consists of hypocellular dermal sclerosing nodules with minimal inflammatory infiltration easily confused clinically and histologically with several dermatoses including annular granuloma, dermatofibroma, or storiform collagenoma. We describe a rare case of EED combining early- and late-stage appearance, characterized by the unusual presentation of nodular sclerotic lesions. Awareness of this peculiar presentation will help avoid misdiagnosis as a neoplastic process in first instance.[2] We recommend performing excisional biopsies whenever possible to avoid misdiagnosis. In addition, early, active lesions that are likely to yield diagnostic results should be preferred.[8]

The present case also highlights the importance of combining therapeutic methods to benefit the patient: dapsone (100 mg/d) to avoid the development of early lesions and surgical excision to treat late-stage lesions.[9] Surgical excision is a treatment option that has been recently described[10] with excellent results and, in the present case, was aimed to relieve discomfort for the patient and prevent interference with usual activities caused by the calcaneal nodules.

ARTICLE INFORMATION

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