

Cutaneous Rosai-Dorfman Disease

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Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy, is a benign disorder of histiocyte proliferation that usually affects the lymph nodes. Cutaneous Rosai-Dorfman disease (CRDD) is a rare extranodal variant that is strictly limited to the skin. We report a patient with CRDD of the trunk. A brief review of the literature of CRDD also is provided. Cutis. 2012;90:237-238.

Case Report

A 78-year-old white man presented with multiple asymptomatic purple lesions on his back of 5 months' duration. On physical examination, 3 indurated, nontender, violaceous nodules were observed on his left flank (Figure 1). No other lesions were noted on the remaining areas of his skin and no lymphadenopathy was appreciated. Systemic examination was unremarkable. He denied any history of fever, chills, night sweats, or weight loss. His medical history was noncontributory and he had no personal or family history of malignancy. His complete blood cell count, complete metabolic panel, and serum protein electrophoresis test were all within reference range. He had not attempted to receive further treatment. A punch biopsy of 1 lesion was performed for further evaluation. Histologic examination of the reticular dermis revealed a proliferation of histiocytes with copious foamy eosinophilic cytoplasm, lymphocytes, and plasma cells (Figure 2). Both large and small histiocytes were present, with several enlarged histiocytes that demonstrated phagocytosis of lymphocytes and plasma cells (emperipolesis). Immunohistochemical staining exposed multiple strongly S-100–positive histiocytes exemplifying emperipolesis of inflammatory

cells (Figure 3). The histiocytes were CD68⁺ and CD1a[−].

Comment

Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy, is a rare benign disorder characterized by the proliferation of histiocytes.¹ Rosai-Dorfman disease most often affects the lymph nodes and more than 40% of patients affected by the disease have extranodal involvement.² Skin involvement is one of the most common extranodal manifestations.³ However, strictly cutaneous disease devoid of systemic involvement has only been reported in 3% of cases.² In the last decade, cutaneous Rosai-Dorfman disease (CRDD) has been recognized as a distinct clinical entity.⁴

Clinically, CRDD has many varieties but often presents with a central noduloplaque surrounded by satellite papules.⁵ It generally affects older white and Asian women, unlike its systemic counterpart, which has a predilection for younger black individuals.⁶ Patients who present with CRDD generally are afebrile and have no systemic concerns. The mean age of presentation is 47 years, and the most commonly



Figure 1. Violaceous nodules on the left flank.

Dr. Parrent is from the University of Kentucky, Lexington. Dr. Clark is from Dermatology Associates of Kentucky, Lexington. Dr. Hall is from Roger Williams Medical Center, Boston University School of Medicine, Massachusetts.

The authors report no conflict of interest.

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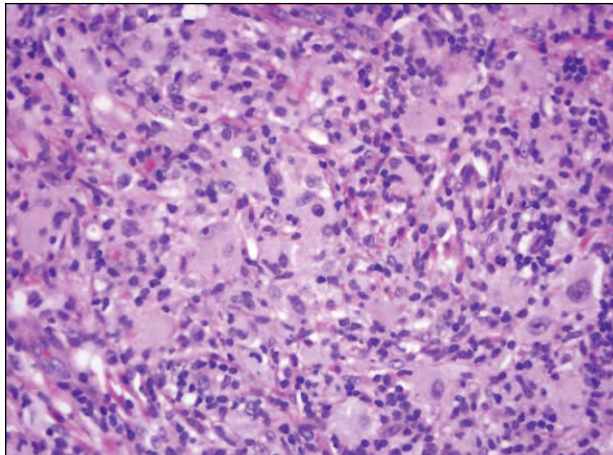


Figure 2. Multiple enlarged histiocytes with some showing engulfed inflammatory cells (H&E, original magnification $\times 40$).

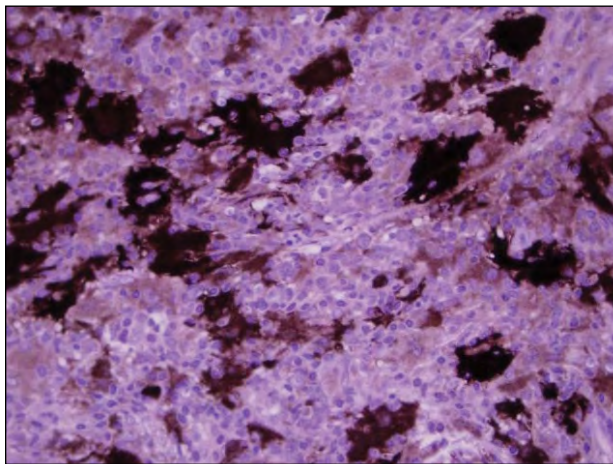


Figure 3. Multiple S-100-positive histiocytes illustrating emperipolesis (original magnification $\times 40$).

affected sites include the extremities, trunk, and face. Laboratory analysis usually does not reveal leukocytosis, anemia, an elevated erythrocyte sedimentation rate, polyclonal hypergammaglobulinemia, or low complement levels.⁷

Histologically, cutaneous lesions may involve the dermis and subcutaneous tissue. The pathognomonic cell is a histiocyte with abundant amorphous pale cytoplasm, indistinct borders, and a large vesicular nucleus containing prominent nucleoli.⁸ These cells can express the macrophage surface markers MAC387 and CD68 as well as the Langerhans cell

marker S-100. Numerous plasma cells, lymphocytes, and polymorphonuclear cells also may be seen on histologic examination.⁸ The hallmark feature of CRDD is the presence of intact inflammatory cells within the cytoplasm of the large histiocytes, a phenomenon known as emperipolesis.⁹

Management of CRDD generally is not necessary, as the disease is benign and usually is self-limited. Surgical excision of cutaneous lesions is effective for unresolved CRDD. Other documented treatments of CRDD include radiation, cryotherapy, chemotherapy, and isotretinoin. Qualification for treatment is dependent on the severity of disease, patient acceptance, and potential complications of therapy.¹⁰

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