Orange Nodules on the Scalp

Marissa L.H. Baranowski, BS; Sarah S. Chisolm, MD; Benjamin K. Stoff, MD; Travis W. Blalock, MD

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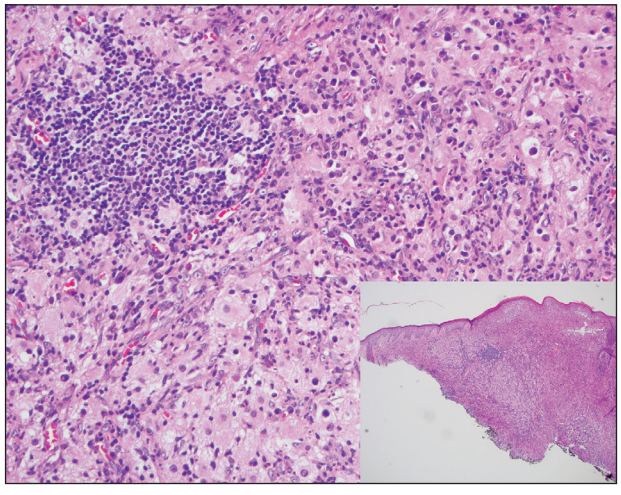

H&E, original magnification ×200 (inset, original magnification ×40).

A 59-year-old man presented with itchy and mildly painful nodules on the head and neck of 7 months' duration. The patient denied fever, chills, unintentional weight loss, night sweats, and other systemic symptoms. Physical examination revealed multiple firm pink-orange nodules of varying sizes distributed on the scalp, face, and neck. Right-sided, painless, bulky cervical lymphadenopathy also was noted. An incisional biopsy was performed.

THE BEST **DIAGNOSIS IS:**

a. blastomycosis
b. granuloma faciale
c. juvenile xanthogranuloma
d. reticulohistiocytoma
e. Rosai-Dorfman disease

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From the Emory University School of Medicine, Atlanta, Georgia. Drs. Chisolm, Stoff, and Blalock are from the Department of Dermatology.
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Correspondence: Benjamin K. Stoff, MD, 1525 Clifton Rd, Atlanta, GA 30329 (bstoff@emory.edu).

THE **DIAGNOSIS:** Rosai-Dorfman Disease

Rosai-Dorfman disease is a rare histiocytic proliferative disorder of unknown etiology. It has 2 forms: limited cutaneous and systemic. The systemic form, also known as sinus histiocytosis with massive lymphadenopathy, affects the lymph nodes and other organs at times. The disease is characterized by a proliferation of histiocytes in the lymph nodes, most commonly in the cervical basin[1]; however, the inguinal, axillary, mediastinal, or para-aortic nodes also may be affected.[1,2] The skin is the most common site of extranodal disease, seen in approximately 10% of cases.[1] Cutaneous involvement often is in the facial area but also can be found on the trunk, ears, neck, arms, legs, and genitals. Clinically, skin lesions appear as papules, plaques, and/or nodules.[2]

Histopathologic examination of Rosai-Dorfman disease generally shows a dense sheetlike dermal infiltrate of large polygonal histiocytes (Figure 1). Histiocytes may display pale pink or clear cytoplasm. The pathognomonic finding is emperipolesis, which consists of histiocytes with engulfed lymphocytes, erythrocytes, plasma cells, and/or granulocytes surrounded by a clear halo. Immunohistochemical staining also is characteristic, with lesional histiocytes showing expression of S-100 protein (Figure 1, inset) and CD68. The associated inflammatory infiltrate is mixed, containing primarily plasma cells but also lymphocytes, neutrophils, and eosinophils.

Blastomycosis (Figure 2) is a systemic infection due to inhalation of *Blastomyces dermatitidis* conidia. Primary infection occurs in the lungs, and with dissemination the skin is the most common subsequently involved organ.[3] Cutaneous blastomycosis shows pseudoepitheliomatous hyperplasia with neutrophilic microabscesses and a dense dermal infiltrate containing suppurative granulomatous inflammation. The nonpigmented yeast phase typically is 8 to 15 μm in length with a refractile cell wall and characteristic single, broad-based budding.[3]

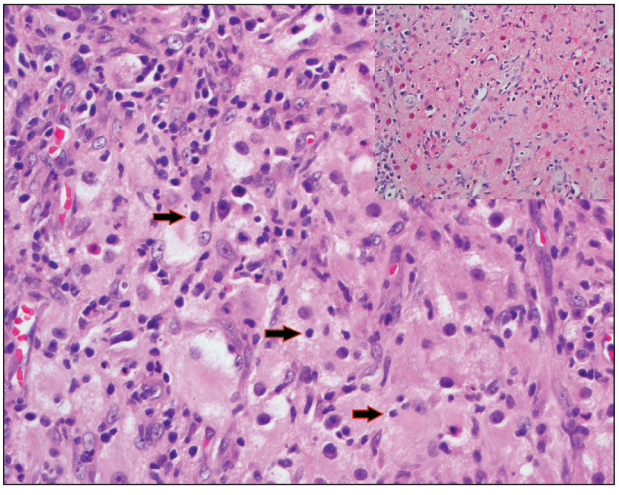

FIGURE 1. Rosai-Dorfman disease showing large polygonal histiocytes and emperipolesis (arrows)(H&E, original magnification ×400). Lesional histiocytes were positive for S-100 protein (inset, original magnification ×400).

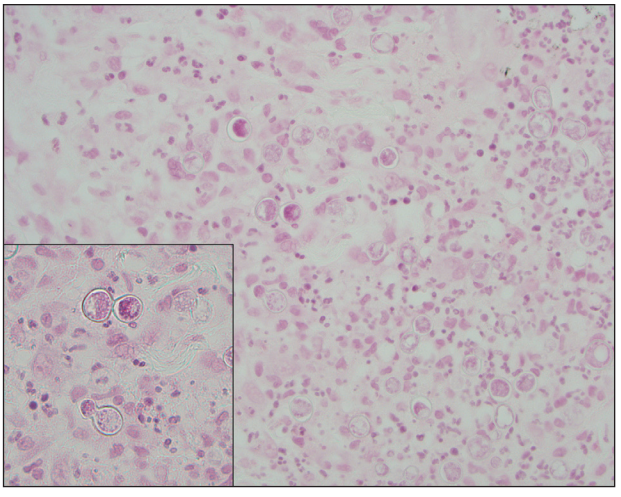

FIGURE 2. Blastomycosis showing a refractile cell wall and broad-based single budding (H&E, original magnification ×400 [inset, original magnification ×400]).

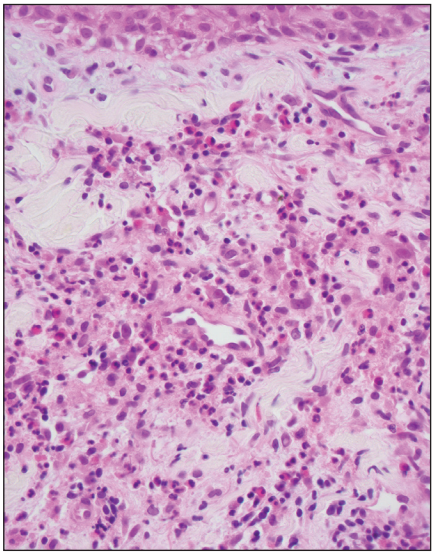

FIGURE 3. Granuloma faciale showing a characteristic grenz zone and a mixed infiltrate of neutrophils with leukocytoclasis and eosinophils (H&E, original magnification ×400).

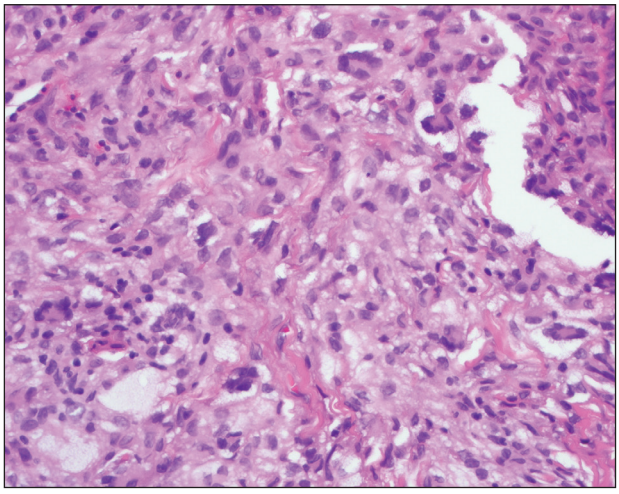

FIGURE 4. Juvenile xanthogranuloma showing foamy histiocytes infiltrating the superficial dermis and characteristic Touton-type multinucleated giant cells with eosinophils (H&E, original magnification ×400).

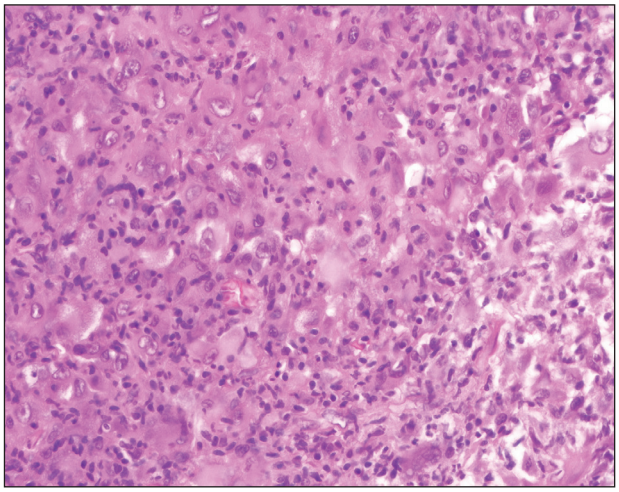

FIGURE 5. Reticulohistiocytoma showing a nodular aggregate of histiocytes with characteristic ground glass granular eosinophilic cytoplasm (H&E, original magnification ×400).

Granuloma faciale (Figure 3) is a rare disease with unknown etiology characterized by reddish brown plaques or nodules most commonly occurring on the face.[4,5] Histology shows a dense nodular dermal infiltrate with a grenz zone. The infiltrate is mixed, containing mostly neutrophils with leukocytoclasis and eosinophils. Leukocytoclastic vasculitis is present with associated extravasated erythrocytes. In chronic fibrosing granuloma faciale, lesions can demonstrate fibrosis and hemosiderin deposition, similar to erythema elevatum diutinum.

Juvenile xanthogranuloma (Figure 4) is a common histiocytic disease of early childhood, though adult cases have been reported.[6] Tumors are found on the head and trunk and are typically firm, reddish yellow papules or nodules.[6,7] Histologic examination shows a nodular infiltrate of foamy histiocytes in the superficial dermis. Touton-type multinucleated giant cells with a peripheral rim of xanthomatized foamy cytoplasm and a wreath-like arrangement of nuclei are characteristic. Associated eosinophils are seen. No emperipolesis is present.

Reticulohistiocytoma (Figure 5) is a benign dermal lesion that presents as solitary or less commonly multiple red-brown papules or nodules.[8] Lesions consist of well-delineated nodular aggregates of histiocytes containing a finely granular eosinophilic ground glass cytoplasm. Few, if any, eosinophils are found. The lack of Touton multinucleated giant cells or emperipolesis and lack of expression of S-100 protein helps to distinguish reticulohistiocytoma from other entities in the differential diagnosis.

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