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Picture Prognosis

A 47-year-old woman presents with a two-year history of red-brown papules, nodules, and comedones on both cheeks. Biopsy shows dermal histiocytes with emperipolesis, plasma cells, fibrosis, and inflammatory infiltrates. Immunohistochemistry is S-100 and CD68 positive and CD1a negative. PET/CT shows only malar soft-tissue involvement without systemic disease. What is the most likely diagnosis?

1. Langerhans cell histiocytosis
2. Primary cutaneous Rosai-Dorfman disease
3. Systemic Rosai-Dorfman disease
4. Cutaneous lymphoma

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Answer: Primary cutaneous Rosai-Dorfman disease

The combination of cutaneous lesions without extracutaneous disease, histiocytic proliferation with emperipolesis, and an immunophenotype of S-100+, CD68+ and CD1a− is characteristic of primary cutaneous Rosai-Dorfman disease. The biopsy showed histiocytes that were S-100 and CD68-positive but CD1a-negative. The absence of CD1a expression argues against Langerhans cell histiocytosis. Classic systemic Rosai-Dorfman disease typically features bilateral painless lymphadenopathy and may involve multiple extranodal organs. In this patient, PET/CT demonstrated disease localized to the malar soft tissues, with no described lymphadenopathy or extracutaneous involvement. The case states that lymphoma was not supported because of the absence of clonality, a mixed inflammatory background, and the absence of atypical lymphoid populations. Instead, characteristic histiocytes with emperipolesis supported Rosai-Dorfman disease.