Isolated cutaneous Rosai-Dorfman disease: a case report.

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Abstract

Background: Rosai-Dorfman also known as sinus histiocytosis with massive lymphadenopathy is a benign, idiopathic lymphoproliferative disorder that usually affects the lymph nodes. Cutaneous Rosai-Dorfman disease is a rare extranodal variant that occurs as histiocyte-rich inflammatory infiltrates, manifesting with a variable clinical morphology. Usually it appears as erythematous to brown papules, plaques, or nodules, with no predilection for site. The histological picture shows abnormal lymph node architecture, reactive germinal centers, fibrosis and emperipolesis in the dermis. On immunophenotypic analysis, S100 protein and CD68 are usually present on dendritic cells.

Case Report: We report a case of purely cutaneous Rosai-Dorfman disease. A 55-year-old male presented to our clinic for an indurated nodule on the left malar region. He reported a slow and progressive growth of 2-year history without systemic symptoms. A cutaneous biopsy showed a nodular inflammatory infiltrate within the dermis consisting of histiocytes, local aggregates of plasma cells and lymphocytes. Histiocytes were enlarged with vesicular nuclei, and emperipolesis was observed. Furthermore, histiocytes stained positively for S-100 and CD68. Owing to local involvement, the patient received a surgery to exsect the lesion completely.

Conclusions: Sinus histiocytosis is a rare inflammatory disease mainly affecting the cervical lymph nodes, presenting with skin lesions in 10% of cases. The diagnosis of cutaneous RDD is differentiated from other histiocytic conditions by the combination of clinical findings accompanied by histopathologic and immunohistochemical confirmation.