

Rosai-Dorfman Disease: A Rare Cause of an Isolated Skin and/or Soft Tissue Mass

Nasir Saleem MD*; Tulsi Kotecha MD; Raya Saba MD

*Nasir Saleem, MD

Department of Internal Medicine, Presence Saint Joseph Hospital, 2900 N Lake Shore Dr, Chicago, IL, 60657, USA. Email: nasirsaleemmd@gmail.com

Description

A 62 year-old Hispanic female presented to the oncology clinic with a right chest wall nodule. She had a past medical history of left-sided breast cancer, diagnosed 37 years prior and treated with left mastectomy and chemotherapy. Her disease recurred in the right breast 8 years later, and was treated with right mastectomy and chemotherapy. She reported having node-positive disease and denied receiving radiotherapy. The patient first noticed the chest wall nodule ten months prior to her current presentation. It was asymptomatic and had not increased in size since. Review of systems was negative; no fever, chills, night sweats or weight loss. Physical examination revealed a 2 cm non-tender mobile nodule on the right chest wall. Complete blood count and comprehensive metabolic panel were within normal limits. Computed tomography of the chest confirmed the presence of a 2.3 cm nodule within the subcutaneous soft tissue of the right lateral chest wall. Metastatic disease was suspected, considering the patient's history, and excisional biopsy of the chest wall nodule was performed. Surgical pathology demonstrated marked sinus histiocytosis intermixed with plasma cells and lymphocytes (Figures 1). Some of the histiocytes exhibited engulfment of lymphocytes, a phenomenon referred to as "emperipolesis" (Figures 2 and 3). Immunohistochemical staining was positive for S-100 and negative for CD1a. The overall findings were consistent with Rosai-Dorfman disease. The patient continued to be in good health and did not have evidence of symptomatic disease or recurrent breast cancer on regular follow up.

Learning Points

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a self-limiting proliferation of non-Langerhans histiocytes that most commonly affects isolated lymph nodes, causing painless lymphadenopathy [1].

Extra-nodal involvement of RDD has been reported in virtually all organ systems, and usually mimics neoplasms. The most common sites of extra-nodal involvement of RDD are the skin and soft tissue [2].

Histologic examination is characterized by numerous large polygonal histiocytes that exhibit abundant eosinophilic cytoplasm. However, the histologic hallmark of RDD is emperipolesis, a phenomenon whereby inflammatory cells such as lymphocytes and plasma cells reside intact within the cytoplasm of histiocytes [3]. Immunohistochemical staining of the histiocytes is characteristically positive for S-100 and CD68 and negative for Cd1a [2].

A high index of suspicion is warranted for the diagnosis of RDD, especially in cases of extra nodal involvement masquerading as malignancy. An accurate histologic diagnosis and appropriate clinical suspicion are crucial to prevent costly diagnostic workup, and resection is curative in most cases.

Figures

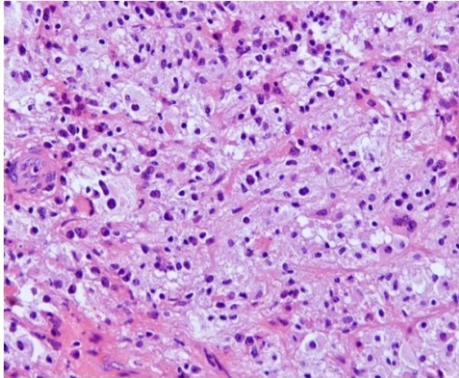


Figure 1: Soft tissue biopsy showing marked histiocytic infiltration (10x, hematoxylin and eosin)

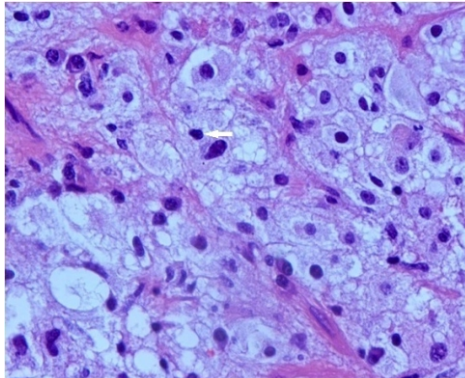


Figure 2: Emperipolesis; an intact lymphocyte within the cytoplasm of histiocyte (white arrow) (40x, hematoxylin and eosin)

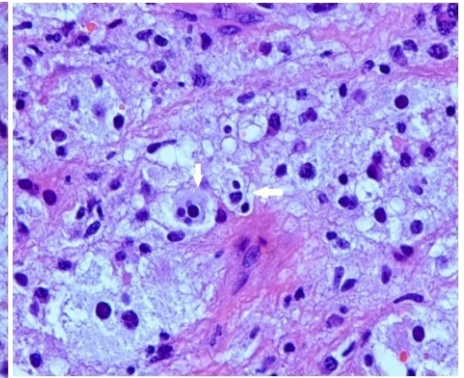


Figure 3: Emperipolesis: lymphocytes within the cytoplasm of histiocyte (white arrows) (40x, hematoxylin and eosin)

References

1. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. *Arch Pathol.* 1969;87(1):63-70.
2. Mantilla JG, Goldberg-Stein S, Wang Y. Extranodal Rosai-Dorfman Disease: Clinicopathologic Series of 10 Patients With Radiologic Correlation and Review of the Literature. *Am J Clin Pathol.* 2016;0:1-11.
3. Chari PS, Iyengar J. Anterior abdominal wall Rosai-Dorfman disease diagnosed by fine-needle aspiration cytology. *J Cytol.* 2015;32(3):214-6.

Manuscript Information: Received: August 05, 2016; Accepted: October 13, 2016; Published: October 15, 2016

Authors Information: Nasir Saleem MD*; Tulsi Kotecha MD; Raya Saba MD
Department of Internal Medicine, Presence Saint Joseph Hospital, 2900 N Lake Shore Dr, Chicago, IL, 60657, USA

Citation: Saleem N, Kotecha T, Saba R. Rosai-Dorfman disease: A rare cause of an isolated skin and/or soft tissue mass. *Open J Clin Med Case Rep.* 2016; 1174

Copy right statement: Content published in the journal follows Creative Commons Attribution License (<http://creativecommons.org/licenses/by/4.0>). © Saleem N 2016

Journal: Open Journal of Clinical and Medical Case Reports is an international, open access, peer reviewed Journal focusing exclusively on case reports covering all areas of clinical & medical sciences.

Visit the journal website at www.jclinmedcasereports.com