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Case Report

Extranodal rosai dorfman disease: a case report of single soft tissue cystic lesion

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ABSTRACT

Rosai-Dorfman disease (RDD) is an uncommon benign histiocytic disorder. It is found worldwide and slightly more common among in men (1.4:1), affecting individuals are with an average age of 20.6 years. Extranodal tissue involvement is documented in 25-43% of RDD patients. Here authors are discussing a case of 16-year-old male patient with the complaint of a single swelling over left forearm since one month with no lymphadenopathy. Radiology showed anechoic collection within the muscle and superficial to it. On microscopy, smear showed diffusely distributed histiocytes with cytoplasm showing intact lymphocytes with halo surrounding it (emperipolesis), without atypia. Based on cytomorphology diagnosis of Extranodal Rosai Dorfman Disease was made.

Keywords: Extranodal rosai dorfman disease, Emperipolesis, Langerhans cell histiocytosis, Rosai dorfman disease, Sinus histiocytosis with massive lymphadenopathy

INTRODUCTION

RDD, is also referred to as sinus histiocytosis with massive lymphadenopathy (SHML), it is a rare non-Langerhans cell reactive histiocytic disorder.[1] The condition was first described in 1965 but only recognized as a clinical and pathological entity in 1969 through a publication by Rosai and Dorfman.[2] RDD or SHML is characterized by non-neoplastic proliferation of histiocytes/phagocytes in the sinusoids of lymph nodes and in extranodal tissues.[3] It belongs to the group of primary histiocytic disorders which includes Langerhans cell histiocytosis and Erdheim-Chester disease and RDD is ultimately differentiated from these disorders based on its characteristic demonstration of emperipolesis, or histiocytic consumption of lymphocytes, on histologic analysis and expression of cellular markers.[4,5]

The objective of this paper was to report a case diagnosed as Extranodal RDD in a cystic swelling over forearm without any lymph node involvement as it is a rare presentation of the Extranodal RDD and also to review the literature.

CASE REPORT

Presenting a case of 16-year-old male patient with the complaint of a single swelling over left forearm since one month. No history of fever, cough, weight loss, night sweats or trauma. On examination patient was well-nourished, afebrile, conscious and well oriented with stable vitals. There was a single swelling over left forearm, measuring 4x3 cm, soft to firm consistency, non-tender, with restricted mobility and overlying skin was normal (Figure 1). There was no lymphadenopathy. Radiology showed anechoic collection within the muscle and superficial to it. Fine needle aspiration was performed, blood tinged fluid was aspirated. On microscopy moderately cellular smear showed diffusely distributed histiocytes with single to multiple nuclei and pale cytoplasm with vacuolation. The cytoplasm of histiocytes showed intact lymphocytes with halo

surrounding it (emperipolesis). Background showed lymphocytes, tingible body macrophages, occasional plasma cells and neutrophils (Figure 2). However, there was no atypia. Based on cytomorphology diagnosis of Extranodal RDD was made.

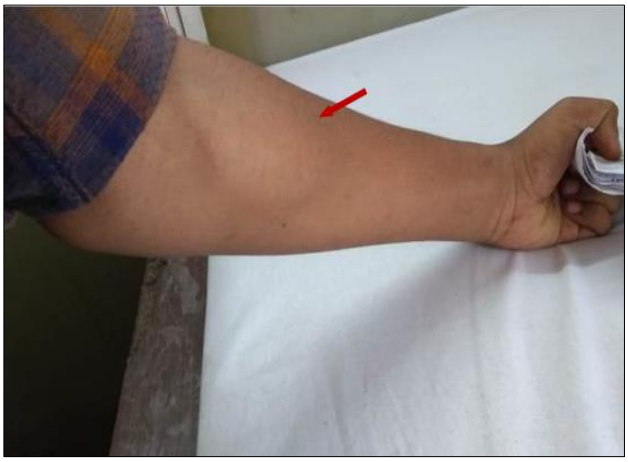

Figure 1: Single swelling over left forearm, measuring 4x3 cm, soft to firm consistency, non-tender, with restricted mobility and overlying skin was normal.

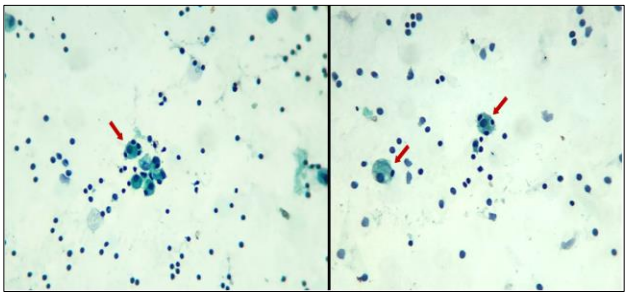

Figure 2: Papanicolaou stain 400x magnification, Smear shows diffusely distributed histiocytes with single to multiple nuclei and pale vacuolated cytoplasm, with intact lymphocytes with halo surrounding it within the cytoplasm (emperipolesis), on background of lymphocytes.

DISCUSSION

Rosai-Dorfman disease (RDD) is an uncommon benign histiocytic disorder.[1] It is found worldwide and slightly more common among men (1.4:1), affecting individuals are with an average age of 20.6 years and is significantly more common among Caucasians and blacks than among Asians.[6] In children it is a rare cause of rapidly progressive lymphadenopathy, which sometimes mimics malignancy.[7] The presentation of disease is typically extensive cervical lymphadenopathy, most often painless and bilateral.[8] Extranodal tissue involvement is documented in 25-43% of RDD patients, especially of the skin, soft tissues, upper airways, oral cavity, urogenital system, bones and lower airways.[9] Extranodal RDD showing involvement of kidney, lower respiratory tract, or liver was found to be a poor prognostic sign, and patients with associated immunologic disease often fared poorly.[8,9] Subcutaneous masses are relatively common and are seen in 9% of RDD cases; therefore, while imaging patients with RDD, adequate attention should be given to the subcutaneous tissue.[1] Subcutaneous masses may extend to the skin surface and presents as a skin lesion. Trunk and proximal extremities are the most common locations. Masses may be solitary or multifocal and are often rapidly growing. [10]

The etiology of RDD is mostly idiopathic the postulated potential sources are autoimmune diseases and viruses such as human herpes virus 6 or Epstein-Barr virus. The pathogenesis of RDD is recruitment of marrow monocytes from the peripheral blood into lymph node sinuses or extranodal sites with subsequent transformation into the immunophenotypically distinct RDD histiocytes.[1] The characteristic feature is emperipolesis of histiocytes in which phagocytized lymphocytes by the histiocytes continue to have free movement in the cytoplasmic vacuoles.[7] Immunohistochemically, RDD shows typical positivity for S-100 and CD68 antigens and negativity for CD1a antigens. However, the important point to note is that CD1a reactivity is more typical for Langerhans cell histiocytosis (LCH) while S-100 reactivity is more characteristic of RDD.[11]

The differential diagnosis of RDD includes lymphoreticular malignancies when cervical lymphadenopathy is present or soft tissue sarcomas, inflammatory myofibroblastic tumor when patients present with extranodal disease. The malignant diagnosis can be dispelled by lack of cytologic atypia, and immunohistochemistry profiles will demonstrate a macrophage-induced histiocytosis with emperipolesis. Distinguishing between RDD-similar S-100-positive histiocytoses such as malignant histiocytosis and LCH is also important. Malignant histiocytosis demonstrates marked cytologic atypia with high mitotic activity, while LCH shows microscopic evidence of Birbeck granules and tends to be CD1a positive. However, the hallmark finding of emperipolesis is absent in both malignant histiocytosis and LCH. The more rare reticulohistiocytoma may be S-100-positive; however, this disease shows prominent ground glass appearance, abundant periodic acid Schiff-positive stain and fewer inflammatory cells in the background. These marker-specific differences are useful in providing a definitive diagnosis.[11]

CONCLUSION

Most cases regress spontaneously, however other modalities for treatment are wide local excision and immunomodulation. To date, surgical resection has proven most successful in preventing recurrences. Patients presenting with suspicious soft tissue masses, cytology helps as a fast and reliable method to

differentiate extranodal soft tissue RDD from its other differential diagnosis. Future studies may help to elucidate the natural history of this disease process, as well as the possibility for malignant potential or transformation to malignancy, thus permitting the development of more evidence-based treatment strategies.

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