

## Case Report

# A case of cutaneous rosai-dorfman disease diagnosed by fine needle aspiration cytology

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### ABSTRACT

A 30 year old female presented with a swelling in the back of a few weeks duration. Clinical examination revealed no other abnormalities. Fine needle aspiration cytology (FNAC) was suggestive of Rosai- Dorfman disease (RDD). The diagnosis was confirmed by biopsy and immunohistochemistry. FNA diagnosis of purely cutaneous RDD has very rarely been reported.

**Keywords:** Rosai-Dorfman disease, Cutaneous, FNAC, Emperipoiesis

### INTRODUCTION

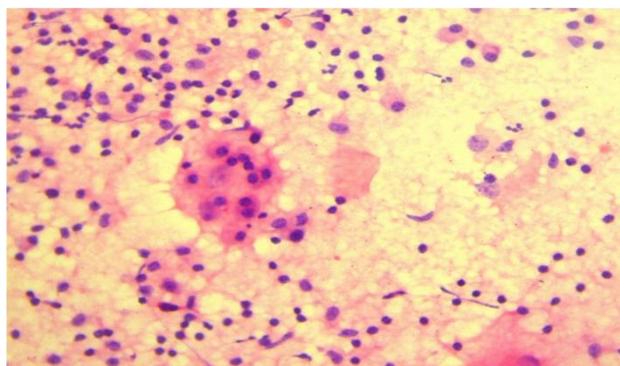
Rosai- Dorfman disease involves extranodal sites in about one third of the cases.<sup>1</sup> 10% are associated with skin and soft tissue involvement.<sup>2</sup> This usually occurs in the presence of massive lymphadenopathy. Extranodal disease as exclusive manifestation is rare.

Herein we report a purely cutaneous Rosai- Dorfman disease without lymphadenopathy initially diagnosed by FNAC.

### CASE REPORT

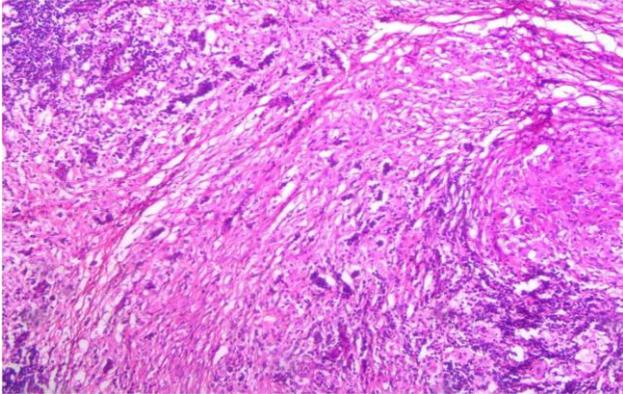
A 32 year old female presented with a swelling in the back of two weeks duration. Examination revealed a hard subcutaneous swelling of size 4x3.5 cm fixed to skin. General and systemic examination was within normal limits. Lab investigations were normal. FNAC was done. Wet fixed smears were stained by haematoxylin and eosin stains (H & E) and air dried smears were stained by May-Grunwald giemsa (MGG) stain. The smears were cellular with sheets of histiocytic cells having abundant

pale cytoplasm and single or multiple bland nuclei with finely dispersed chromatin. Some of these cells showed ingested intact lymphocytes and plasma cells (emperipoiesis). Background showed numerous foam cells, plasma cells and lymphocytes (Figure 1). With these findings, the diagnosis of Rosai- Dorfman disease was suggested. Subsequently excision of the lesion was done.

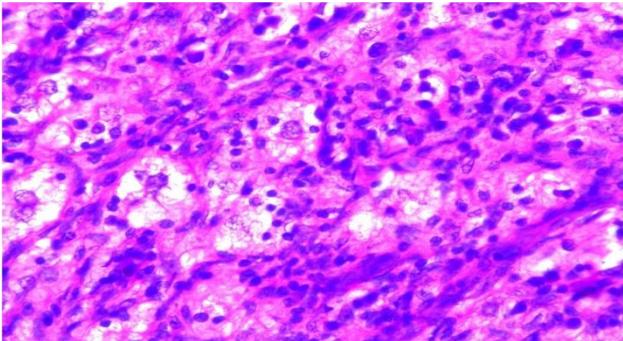


**Figure 1: FNA smear showing histiocytes with emperipoiesis and lymphocytes (H&E, 400x).**

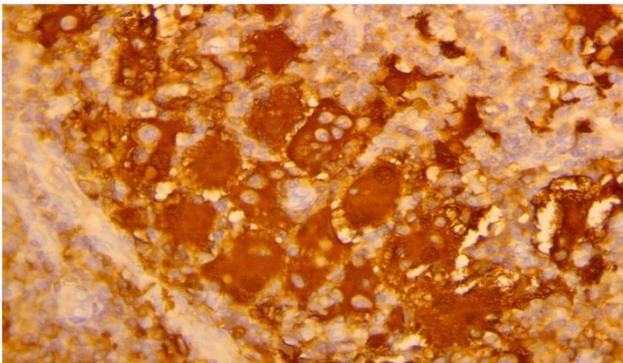
The specimen received was multiple irregular pieces of firm tissue aggregate measuring 4.5x3.5x3 cm with lobulated white and yellow cut surface. Microscopy showed sheets of mixed inflammatory infiltrate including lymphocytes, plasma cells and multinucleated histiocytes separated into nodules by bands of fibrous tissue (Figure 2). Large aggregates of pale staining histiocytes demonstrating emperipolesis was seen as shown in Figure 3. Immunohistochemical stain for S100 was strongly positive in the histiocytes (Figure 4). The histologic and immunohistochemical findings were consistent with RDD.



**Figure 2: Histology showing mixed infiltrate and foamy histiocytes (H&E, 400X).**



**Figure 3: Histology showing emperipolesis (H&E, 400X).**



**Figure 4: The cells are positive for S100 (1000X).**

## DISCUSSION

Rosai-Dorfman disease or sinus histiocytosis with massive lymphadenopathy (SHML) is a rare benign disorder of histiocytes characterized histologically by emperipolesis.

SHML was initially described by Destombes in 1965.<sup>3</sup> Rosai and Dorfman coined the term sinus histiocytosis with massive lymphadenopathy and described this disorder as massive bilateral and mostly cervical lymphadenopathy in young black children.<sup>4,5</sup> A review of 423 cases published in 1990 revealed a much broader spectrum of disease with wide geographic distribution.<sup>1</sup> The age of patients range from newborn to 74 years (mean, 20.6 years). Males are more commonly affected than females.<sup>1</sup> In addition to lymph nodes, in approximately one-third of patients, SHML, can occur in a variety of extranodal sites.<sup>1</sup> Virtually any extranodal site can be affected, but the head and neck region is most common.<sup>6</sup> Other commonly involved extranodal sites include the soft tissue, skin, upper respiratory tract, gastrointestinal tract, breast, bones, and the central nervous system.<sup>7-12</sup> About 10% of patients with nodal disease have cutaneous involvement also.<sup>2</sup> The lesions are in the form of papules or nodules. The skin and subcutaneous soft tissue are common sites of involvement when patients present with extranodal disease without nodal involvement.<sup>13,14</sup> Some consider cutaneous RDD as a distinct entity related to nodal RDD. Many of these do not progress to involve lymph nodes or other systemic sites.<sup>15</sup>

### Cytology

Fine needle aspiration (FNA) smears and touch imprints are usually cellular with many histiocytes and phagocytosed lymphocytes in a reactive background of inflammatory cells.<sup>16,17</sup> The histiocytes have abundant, pale, sometimes vacuolated cytoplasm. The background inflammatory cells are predominantly lymphocytes in early stages whereas; plasma cells predominate in later stages.<sup>17,18</sup>

In cytology preparations, a halo surrounding phagocytosed lymphocytes are not seen as seen in tissue sections which is a result of fixation artifact.<sup>3</sup> This can lead to difficulty in distinguishing emperipolesis from overlying lymphocytes. The diagnosis can be missed if the index of suspicion is not high. Only a few well documented cases diagnosed by FNAC have been reported.<sup>19</sup> FNA diagnosis of pure cutaneous RDD is rather difficult as clinical findings are not contributory.

### Histopathology

The skin lesions contain a mixed infiltrate in which lymphocytes, plasma cells and macrophages with clear cytoplasm are the most prominent. Occasionally the macrophages may be multinucleated or have a foamy

cytoplasm. The hallmark histologic feature is emperipolesis of lymphocytes.<sup>20</sup> Emperipolesis differs from phagocytosis in that the lymphocytes are taken up intact. On occasion, red cells can also be taken up. Cutaneous lesions differ histologically from nodal diseases in that there is a greater degree of fibrosis, less number of histiocytes and reduced emperipolesis. Differential diagnosis includes a variety of histiocytic lesions and lymphomas.

### Immunohistochemistry

SHML cells belong to the macrophage-histiocyte family and are positive for the monocyte/macrophage-associated antigens. In most cases, they also stain with antibodies to  $\alpha$ 1-antichymotrypsin and  $\alpha$ 1-antitrypsin, which suggests lysosomal activity. In almost all cases, SHML cells also express S100 protein. S100 protein is also expressed by interdigitating dendritic cells in lymph nodes and Langerhans cells in skin. However, SHML cells are morphologically distinct from either interdigitating dendritic cells or Langerhans cells. CD1a is not detected in SHML cells but is positive with Langerhans cells.<sup>21</sup>

RDD presenting exclusively as extranodal disease is rare and a high index of suspicion is necessary for diagnosis. It should be considered in the differential diagnosis of all cutaneous and soft tissue lesions with histiocytic proliferation. Emperipolesis is the key feature for diagnosis. Cytologic diagnosis helps in differentiation from sarcomas thereby preventing radical surgery.

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