

## Soft Tissue Rosai-Dorfman Disease in a 77-Year-Old Female

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### Abstract

Rosai-Dorfman disease is a histiocytic disorder that is most common in children and young adults of African descent which typically presents as painless massive cervical lymphadenopathy. The purpose of this article is to report a case of Rosai-Dorfman disease in a 77-year-old woman who presented with a large subcutaneous mass in her lower left abdomen. Microscopic examination revealed diagnostic features of Rosa-Dorfman disease. This uncommon disorder may involve lymph nodes and other organs, but rarely involves and presents as a soft tissue tumor. Clinical observation is the usual treatment, and correct diagnosis is important to avoid unnecessary therapy.

**Keywords:** Histiocytosis; Lymphadenopathy; Emperipolesis; Rosai-Dorfman disease

**Abbreviations:** RDD: Rosai-Dorfman Disease; AAF: African American Female; EBV: Epstein Bar Virus; HHV: Human Herpes Virus; SV40: Simian Virus 40; Hct: hematocrit; Hgb: Hemoglobin; CBC: Complete Blood Count; CT: Computed Tomography; CD: Cluster of Differentiation; INR: International Normalized Ratio; PT: Protime

### Introduction

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare disorder first described by Rosai and Dorfman in 1969. Rosai and Dorfman analyzed four cases of lymphadenopathy that were found to have the consistent feature of phagocytized lymphocytes known as emperipolesis [1]. The definite etiology of the disease is not well understood; several viruses including HHV-6, rubella, and EBV have been proposed to play a part in the pathogenesis [2]. Recently, simian virus 40 (SV40) has been postulated to play a role in the pathogenesis of RDD [3]. RDD most often occurs in children and young adults of African descent with a male predominance. The disease usually presents with nonspecific findings such as anemia, polyclonal hypergammaglobulinemia, fever, thrombocytosis, and lymphadenopathy that tends to occur in the head and neck [4]. Cervical lymph nodes are most commonly affected; however, other lymph nodes as well as extra nodal sites may be involved [5].

Here, we report a case of RDD presenting as a soft tissue mass in an elderly female.

### Case Report

A 77-year-old African American female presented to the Gastroenterology Department on March 2015 with a history of microcytic anemia. Past medical history was remarkable for recurrent deep vein thromboses, hypertension, herpes zoster (shingles), asthma and disseminated tuberculosis successfully treated. Medications included coumadin, lisinopril-hydrochlorothiazide, and fluticasone. Endoscopy revealed no source of bleeding, but during the abdominal physical exam, an approximately 2-inch non-tender, hard, movable mass was identified in the patient's lower left quadrant. A CBC with differential showed Hct of 28.2% (normal 35 - 47%) and Hgb 9.4 g/dl (normal 12.0 - 16.0 g/dL). PT was 19.2 seconds (normal 9.3 - 11.1 seconds) with an INR of 2 (therapeutic range 2 - 3). The CT of the abdomen and pelvis revealed an 8.7 × 4.0 × 7.3 centimeter mass involving the subcutaneous fat with a clear fat plane separating the mass from the underlying muscle (Figure 1). An ultrasound guided needle biopsy of the mass was obtained and sent for histopathological examination.



**Figure 1:** CT scan reveals a large soft tissue mass like within the left lower anterior abdominal wall involving the subcutaneous fat with a clear fat plane from the underlying intra-abdominal musculature. The mass measures approximately 8.7 × 4.0 × 7.3 cm.

### Pathologic Findings

Microscopic examination showed collagenous tissue infiltrated by polygonal to spindled histiocytoid cells with clear to pale eosinophilic vacuolated cytoplasm. Nuclei showed irregular, indented contours. Nucleoli were not prominent, and mitoses were not evident. Rare histiocytoid cells displayed intracytoplasmic lymphocytes. Scattered plasma cells and collections of lymphocytes were present (Figure 2). Immunoperoxidase stains were performed and the polygonal, histiocytoid cells showed negative staining for SV40, strong positive cytoplasmic staining in polygonal cells with clear to vesicular cytoplasm for CD-68, perivascular positive staining for actin and desmin, and strong positive S-100 nuclear and cytoplasmic staining of histiocytoid cells (Figure 3). Based on the morphological and immunohistochemical findings, the diagnosis of soft tissue Rosai-Dorfman disease was made.

### Clinical Follow-Up

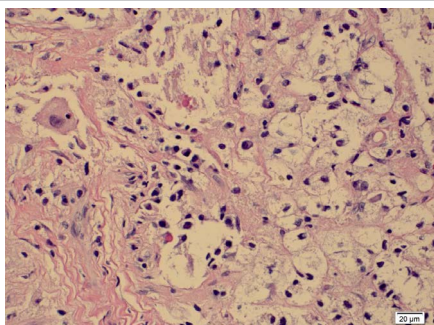
The patient was sent home following biopsy with instructions to

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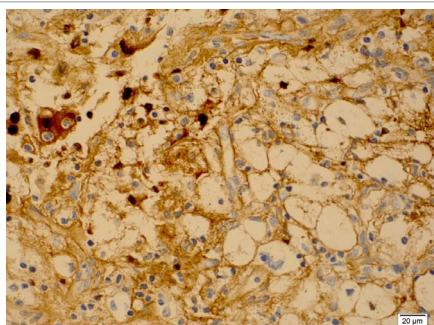
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**Figure 2:** Hematoxylin-and-eosin stained sections show histiocytoid cells with clear to pale eosinophilic vacuolated cytoplasm. Some nuclei appear indented and resemble lipoblasts. Plasma cells, lymphocytes, and histiocytoid cells with intracytoplasmic lymphocytes are present.



**Figure 3:** An S-100 immunostain demonstrates cytoplasmic and nuclear positivity in histiocytoid cells, and helps identify cells with intracytoplasmic lymphocytes.

limit activity and avoid non-steroidal anti-inflammatory drugs for 48 hours. She is being followed clinically and has had no complaints related to her abdominal mass.

## Discussion

As described by Rosai and Dorfman in 1969, RDD is a rare disorder with distinct clinical and pathological findings. It most commonly affects male children and young adults, and often presents as painless massive cervical lymphadenopathy, fever, neutrophilia, elevated ESR, and hypergammaglobulinemia. It is interesting to note that soft tissue RDD may not have this typical presentation, with female predominance and older age at presentation [6], as was seen in our case.

Common histological findings of RDD in lymph nodes include: pericapsular fibrosis, dilation of sinuses, histiocytes with abundant clear to vesicular cytoplasm, and the distinctive finding of cells showing emperipolesis [7]. Immunohistochemical stains are useful in the diagnosis of SHML. In particular, the histiocytoid cells show positivity for CD-68 and S-100 protein and are negative for CD-1a [8].

Frequently, the natural course of the disease is protracted; however, the majority of patients will have symptoms lasting several years with spontaneous resolution. Nevertheless, a study that reviewed 215 cases found that RDD was either the direct or indirect cause of death in 14 patients [9]. Although direct infiltration can be a cause of mortality, immune dysfunction is thought to carry the greatest risk in patients with RDD [10]. In our patient, the history of herpes zoster (shingles) and disseminated tuberculosis is consistent with some level of immune dysfunction. Patients with RDD should be monitored for unusual or opportunistic infections.

Extra nodal infiltration of sites that include testes, kidney, spleen,

CNS, liver, bone, eye-lid, orbit, respiratory tract, skin, and salivary glands has been reported in previous case reports [11]. This patient presented with an incidental large painless abdominal wall mass on physical examination, and the clinical differential diagnosis included benign and malignant soft tissue neoplasms as well as inflammatory, non-neoplastic, soft tissue proliferations. Definitive diagnosis requires biopsy and pathologic examination. Pathologic diagnosis may be difficult, especially when diagnostic material submitted for pathologic examination consists of limited needle core biopsies. In one review of 23 soft tissue lesions from 17 patients, soft tissue RDD was initially correctly diagnosed in only one patient [6]. The morphologic features of soft tissue RDD, in particular the relative rarity of cells with the characteristic finding of emperipolesis, and its collagenous background and associated inflammatory infiltrate, make correct diagnosis difficult. These atypical morphologic features may suggest a diagnosis of inflammatory pseudotumor or inflammatory sarcoma. In our patient, the finding of polygonal cells with vacuolated clear to pale eosinophilic cytoplasm with indented nuclei was felt to be suggestive of lipoblastic differentiation. Recognition of the cells as histiocytoid rather than lipoblastic, along with the finding of emperipolesis was critical for determining the correct diagnosis. must be considered in the differential diagnosis of benign and malignant inflammatory soft tissue tumors.

## Conclusions

In summary, soft tissue RDD is a rare disorder, which most often behaves in a benign manner and follows an indolent course. In order to avoid unnecessary and potentially harmful therapy, RDD must not only be included in the differential diagnosis of painless cervical lymphadenopathy in the characteristic clinical setting of a male child or young adult children and young adults with fever, neutrophilia, elevated ESR, and hypergammaglobulinemia, but in the differential diagnosis of soft tissue tumors in older individuals regardless of gender.

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