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Imad ud Deen

*Medstar Health Internal Medicine Residency Program, imadud.deen@medstar.net*

Abhinandan Chittal

*Medstar Health Internal Medicine Residency Program*

Nabil Badro

*Department of Surgery, Medstar Health, Harbor Hospital*

Robert Jones

*Department of Pathology, Medstar Health*

Christopher Haas

*Department of Medicine, Medstar Health*

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# Extranodal Rosai-Dorfman Disease- a Review of Diagnostic Testing and Management

Imad ud Deen <sup>a,\*</sup>, Abhinandan Chittal <sup>a</sup>, Nabil Badro <sup>b</sup>, Robert Jones <sup>c</sup>, Christopher Haas <sup>d,e</sup>

<sup>a</sup> Medstar Health Internal Medicine Residency Program, USA

<sup>b</sup> Department of Surgery, Medstar Health, Harbor Hospital, USA

<sup>c</sup> Department of Pathology, Medstar Health, USA

<sup>d</sup> Department of Medicine, Medstar Health, USA

<sup>e</sup> Department of Medicine, Georgetown University Medical Center, USA

## Abstract

Rosai-Dorfman disease (RDD) is a rare, benign histiocytic proliferative disorder primarily occurring in the lymph nodes. Extranodal disease can occur in skin and soft tissue, central nervous system, gastrointestinal tract, and breast. Here we present a case of a 55-year-old female with a history of hypertension, hyperlipidemia, and diabetes mellitus who presented with a fixed solid mass localized to the right medial thigh. Excision revealed S100 and CD 68 positive scattered lymphoid aggregates and emperipolesis, hallmark molecular and cytopathologic features of RDD. RDD is a nonmalignant, classically sporadic histiocytosis. Clinical outcome in terms of mortality seems to be favorable in most cases, however given the benign and slowly progressive nature of the disease, conservative management with observation is typical, with surgical excision recommended for larger lesions. Currently, therapy revolves around surgical and immunosuppressive treatments, but the optimal diagnostic and therapeutic management of RDD remains to be defined.

**Keywords:** Rosai-Dorfman disease

## 1. Background

Rosai-Dorfman disease (RDD), first described as a distinct clinicopathological entity in 1969, is a rare non-malignant disorder characterized by histiocyte proliferation with a prevalence of only 1 in 200,000 in the United States.<sup>1,2</sup> The disease predominantly affects children and young adults, with a slight male predominance, and oftentimes displays a chronic, relapsing course.<sup>3,4</sup> Patients typically present with fever, leukocytosis, and painless cervical lymphadenopathy. Although RDD has a predilection for the lymph nodes of the head and neck, any nodal region may be affected, and furthermore the disease may also present in extranodal locations including the skin and soft tissue, central nervous system (CNS), gastrointestinal tract, and breast.<sup>5–12</sup> In contrast to nodal RDD, extranodal, cutaneous RDD usually presents as slow-growing, painless, nonpruritic nodules, plaques, or

papules with coloration varying from yellow to red to brown.<sup>13</sup> Emperipolesis, histiocytic phagocytosis of lymphocytes, plasma cells, erythrocytes, or polymorphonuclear leukocyte, is considered to be pathognomonic and remains a critical histopathological feature, along with S-100 and CD68 positive staining of the histiophagocytic cells.<sup>1,14–16</sup> We present a rare case of Rosai-Dorfman disease, an underrecognized and diagnosed entity with limited, albeit emerging diagnostic criteria, heretofore focused on immunohistochemistry of tissue samples. We also explore management options, highlighting both conservative evaluation as well as surgical intervention with periodic surveillance for resolution or recurrence.

## 2. Case presentation

A 55-year-old woman with a past medical history significant for hypertension, hyperlipidemia, non-insulin dependent diabetes mellitus, and right

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\* Corresponding author.  
E-mail address: [imadud.deen@medstar.net](mailto:imadud.deen@medstar.net) (I.ud Deen).

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middle cerebral artery infarction managed with mechanical thrombectomy presented with complaints of right thigh swelling. She reported usual health until six months prior to presentation when she noticed a solid mass on the medial aspect of her right thigh. There was no associated pain, redness, or discharge, and no antecedent trauma. Physical examination demonstrated an 11 × 6 cm fixed solid mass localized to the right medial thigh without erythema, crepitus, or ecchymoses. Diagnostic evaluation demonstrated unremarkable serum chemistries. She underwent surgical resection of the mass with gross examination notable for a tan-yellow lesion measuring 6.1 × 4.8 × 2.5 cm surrounded by normal subcutaneous tissue. Tissue pathology demonstrated diffuse, sheet-like proliferation of spindled and epithelioid cells in a background of sclerotic and fibrotic stroma (Fig. 1). Histological analysis revealed involvement of mature adipose tissue and chronic inflammation with the presence of scattered lymphoid aggregates and plasma cells along with histiocytes containing ample eosinophilic granular cytoplasm and intact intracytoplasmic inflammatory cells (“emperipolesis”) (Fig. 2). Immunohistochemical staining was negative for CD34, actin, desmin, beta-catenin, CD1a, and mycobacterial/fungal stains, but notably positive for S100 (Fig. 3) and CD68, further confirming the diagnosis. The patient's wound was, while large, was closed by primary intention, without tension. She was followed weekly on an outpatient basis with appropriate approximation and closure. She was followed

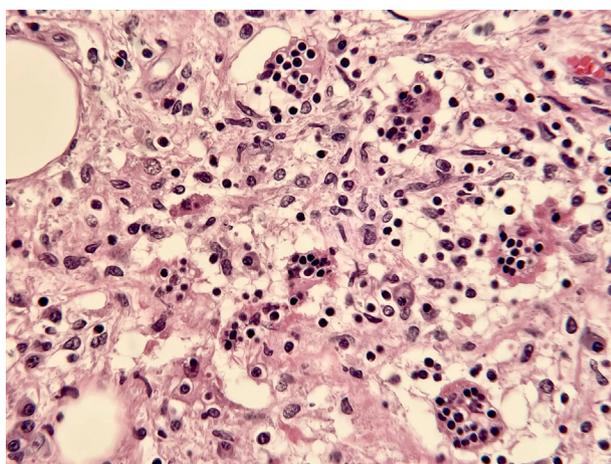


Fig. 1. Hematoxylin and Eosin staining. At low power magnification, sheets and syncytial clusters of histiocytes with abundant pink cytoplasm are admixed with subcutaneous fat (clear spaces, top left and bottom left corners). Histiocytes containing numerous lymphocytes can be seen within the cytoplasm of the histiocytes (right side of image), a histopathologic feature known as “emperipolesis,” the presence of an intact cell within the cytoplasm of another cell.

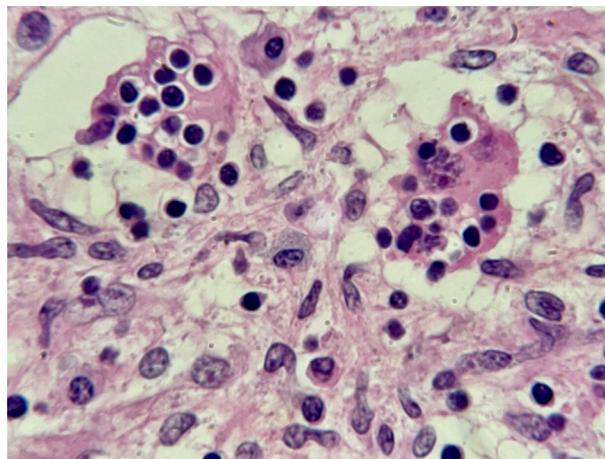


Fig. 2. Hematoxylin and Eosin staining. At higher power, emperipolesis can be better appreciated. This image contains two histiocytes with numerous lymphocytes (seen as blue circles with clear halos in this image) passing through their pink cytoplasm. Emperipolesis, when seen, is pathognomonic for an underlying diagnosis of Rosai-Dorfman Disease.

for 6 months, at the time of this report, with no evidence of local recurrence or dehiscence.

### 3. Discussion

Rosai-Dorfman disease is a rare, benign, idiopathic proliferative disorder affecting histiocytes,<sup>1</sup> typically affecting isolated lymph nodes.<sup>17–19</sup> Patients present with bilateral, painless, cervical lymphadenopathy with or without constitutional symptoms such as fever, night sweats, and weight loss. Although classically confined to lymph nodes, up to 40% of patients with RDD manifest extra nodal involvement with or

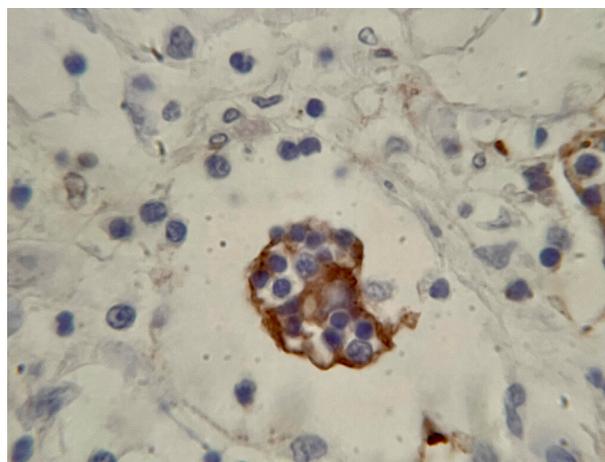


Fig. 3. S-100 Immunostaining. S-100 is an immunostain that is positive in histiocytes and can help highlight emperipolesis. Here, we see S-100 highlighting the cell membrane of the histiocytes, which is negative in the cytoplasmic lymphocytes, another classic histologic finding in Rosai-Dorfman Disease.

without adenopathy. In contrast to nodal disease, extranodal disease most commonly presents as a painless, palpable mass. It affects females more frequently, with one case series documenting a 90% female predominance.<sup>20,21</sup> The most frequent extranodal sites were found to be skin and soft tissue (16%); nasal cavity and paranasal sinuses (16%); eye, orbit, and ocular adnexa (11%); bone (11%); salivary gland (7%); central nervous system (7%); oral cavity (4%); kidney and genitourinary tract (3%); respiratory tract (3%); liver (1%); tonsil (1%); and breast (<1%).<sup>17</sup> Our patient's presentation correlates well with previously reported cases of extranodal RDD, presenting with an insidious, slowly progressing, asymptomatic mass. While the majority of RDD is considered to be nodal in nature, an extensive physical evaluation was performed, yet failed to demonstrate any evidence of lymphatic swelling. While some patients with RDD have adjunctive constitutional symptoms – fever, night sweats, weight loss – this is not considered to be exclusive and may raise the specter of an alternative diagnosis such as a lymphoproliferative disease. Our patient did not experience any of these symptoms and our suspicion for an alternative diagnosis, prior to histological sampling, remained low.

The presence of nonspecific symptoms such as fever, leukocytosis, weight loss, and cervical lymphadenopathy in individuals with RDD may mimic a variety of lymphoproliferative disorders: Hodgkin's and non-Hodgkin's lymphoma, Langerhans cell histiocytosis, granulomatous lymphadenitis, reactive lymphadenopathy, Castleman's Disease, and Kikuchi-Fujimoto disease – making histopathology a key feature of diagnosis.<sup>22</sup> Histologically, RDD is characterized not only by histiocytic proliferation, but also histiocyte-mediated phagocytosis of intact lymphocytes and other immune cells, leading to the disease's histological hallmark finding, emperipolesis.<sup>23</sup> These histiocytes are typically positive for S-100 and CD68 antigens and negative for CD1a immunohistochemical staining and BRAF V600E mutations.<sup>24,25</sup> These findings are used to differentiate RDD from Langerhans's cell histiocytosis and Erdheim-Chester Disease, in which the histopathological findings are characterized by a lack of emperipolesis and the presence of Langerhans' cells and foamy histiocytes.<sup>26–28</sup> Furthermore, these disorders appear to have unique immunohistochemical signatures: S100-negative, CD68-positive, CD1a negative, and BRAF V600E positive cells in Erdheim-Chester; S100-positive, CD68-positive, CD1a positive, and BRAF V600E positive cells in Langerhans's cell histiocytosis.

The pathogenesis of RDD remains unknown. Infection by human herpes virus-6 (HHV-6), Epstein–Barr virus (EBV), and Parvovirus B19 have been suspected as instigating events.<sup>29–32</sup> Several genetic mutations have also been identified recently including mutations in *ARAF*,<sup>33</sup> *MAP2K1*,<sup>34,35</sup> *NRAS*, and *KRAS*.<sup>33</sup> *KRAS* and *MAP2K1* are associated with 33% of RDD cases, suggesting that the RAS-MAP2K1 pathway may be an integral component of disease development and thus a potential therapeutic target.<sup>36</sup>

Of note, our patient had an initial breast biopsy demonstrating CD68-positive cells, which while not specific, could have raised suspicion for extranodal, breast RDD, specifically if additional staining for S-100 (positive) and the absence of a BRAF V600E mutation were noted. However, at the time she was diagnosed with benign histiocytic infiltration without further follow up. While likely unrelated, RDD of the breast may mimic fat necrosis on core biopsy.<sup>37</sup> She subsequently presented with soft tissue swelling on the medial aspect of the thigh, which was removed surgically with subsequent histopathological findings consistent with RDD. Review of cases with RDD involving the CNS show a 15% recurrence rate after surgery,<sup>38</sup> though no literature has been reported regarding recurrence of cutaneous RDD.

The diagnostic and staging evaluation of patients with newly diagnosed RDD should include an assessment of disease extent, as well as evaluation for conditions either known to be associated with RDD, particularly autoimmune disorders, or known to contain a component of reactive histiocytosis.<sup>39</sup> CT of the neck, chest, abdomen, and pelvis is recommended. RDD lesions are known to be FDG-avid, including extra nodal areas<sup>40</sup> and FDG-PET/CT is used by some investigators for initial staging when possible.<sup>41</sup> Biopsy is required for confirmatory diagnosis.

RDD resolves spontaneously in approximately 20%–50% of the cases,<sup>29,42</sup> however a minority of patients may develop organ dysfunction due to massive nodal enlargement resulting in fatal disease. Treatment is warranted to halt the natural progression of RDD in such cases. Therapeutic options for RDD include corticosteroid therapy, surgery, radiation therapy and biologics.<sup>44</sup> The natural history of the disease is uncertain, however, the reported mortality rate is about 7%; with concomitant presence of immune dysfunction.<sup>42–44</sup> Emerging data suggests that given the low rate of mortality, even in the absence of intervention, more patients are electing conservative management with observation as opposed to surgical excision<sup>45</sup>;

however given that our patient had a large mass, surgical excision was the treatment of choice.

#### 4. Conclusion

Rosai-Dorfman Disease is a rare, benign histoproliferative disease mainly affecting the lymph nodes of the head and neck, however the disease may manifest in alternative lymphatic groups as well as extranodal sites, including the skin. Confirmatory diagnosis is done by biopsy with histopathological and immunostaining, with classic demonstration of histiocytic proliferation, histiocyte-mediated phagocytosis of intact lymphocytes and other immune cells, and positive staining for S-100 and CD68 antigens. While the disease may resolve spontaneously, surgical excision is recommended for larger tumor burden.

#### Declaration of competing interest

There is no conflict of interest.

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