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

Edgar Zamora* and Peter Kragel

1Brody School of Medicine, East Carolina University, USA
2Department of Pathology, Brody School of Medicine, East Carolina University, USA

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 Rossa-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 know 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 role 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 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 nontender, 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.

Pathologic Findings

Microscopic examination showed collagenous tissue infiltrated by polygonal 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

*Corresponding author: Peter Kragel, Department of Pathology, Brody School of Medicine, East Carolina University, USA, Tel: 252-847-4951; Fax: 252-847-6368; E-mail: kragelp@ecu.edu

Received December 03, 2015; Accepted January 05, 2016; Published January 09, 2016


Copyright: © 2016 Zamora E, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.
opportunistic infections. With RDD [10]. In our patient, the history of herpes zoster (shingles)
immune dysfunction is thought to carry the greatest risk in patients [9]. Although direct infiltration can be a cause of mortality,
found that RDD was either the direct or indirect cause of death in 14
the majority of patients will have symptoms lasting several years with
Figure 2: Hematoxylin-and-eosin stained sections show histiocytoid cells with
clear to vesicular 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.

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 central
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.

References
recognized benign clinicopathological entity. Archives of Pathology 87: 63-70.
detection of polyomavirus antigen in 3 abdominal cases. Annals of Diagnostic
Pathology 14: 309-316.
multi-organ involvement in head and neck region. Int J Pediatr Otorhinolaryngol
68: 581-584.
8. Ahsan SF, Madgyn DY, Poulik J (2001) Otolaryngologic manifestations of
59: 221-227.
press 586-587.