Available online at www.sciencedirect.com

ScienceDirect

journal homepage: http://Elsevier.com/locate/radcr

Case Report

# Extranodal manifestation of Rosai-Dorfman disease in the breast tissue

Qiao Zhou MD[a,*], Umer Ansari BS[a], Nandan Keshav BS[a], Fiona Davis MD[b], Maria Cundiff MD[b]

[a] Department of Radiology, Eastern Virginia Medical School, 6800 Gresham Dr Norfolk, VA 23510, USA
[b] Department of Radiology, Sentara Norfolk General Hospital, Norfolk, VA, USA

ARTICLE INFO

*Article history:*
Received 14 December 2015
Received in revised form
24 January 2016
Accepted 17 April 2016
Available online 17 May 2016

*Keywords:*
Rosai Dorfman disease
Mammography
Histiocytes
Breast

ABSTRACT

A 71-year-old asymptomatic female with a history of breast cancer status after right total mastectomy had interval development of several new nodules in the left breast in a 1-year time span. Stereotactic biopsy was performed, which revealed multifocal Rosai Dorfman disease in the left breast. The patient was referred to hematology, and computed tomography of the chest/abdomen/pelvis did not demonstrate any lymphadenopathy elsewhere in the body. This case report discusses incidences of extranodal Rosai Dorfman disease and the differential for breast lesions that can present the same way.



## Case report

A 71-year-old woman with a history of breast cancer s/p right total mastectomy, hypertension, hypothyroidism, and right knee osteoarthritis presented to the clinic for a regularly scheduled annual screening mammogram. On comparison with previous mammograms (Fig. 1), several new nodules were noted in the left breast (Fig. 2). Diagnostic mammography confirmed the persistence of 2 new nodules within the left breast. Ultrasonography confirmed the presence of an oval parallel $5 \times 3 \times 5$ $mm^3$ mass with slightly irregular margins at the 12:00 position, as well as a $4 \times 4 \times 3$ $mm^3$ hypoechoic nodule near the 2:00 position (Fig. 3). A Breast Imaging Reporting and Data System of 4 was given, and biopsy of these nodules was recommended.

A stereotactic core biopsy of the left breast was chosen over ultrasound guidance because of close proximity of these nodules, and a total of 9 core samples were obtained. The pathology of these nodules was later found to be extranodal Rosai-Dorfman disease (RDD; Figs. 4 and 5). This finding was deemed benign, but further evaluation for additional involvement of nonbreast areas was recommended, and patient follow-up with the hematology service was advised as well. Subsequent computed tomography (CT) of the chest abdomen pelvis did not demonstrate any evidence of

Competing Interests: The authors have declared that no competing interests exist.

* *Corresponding author.*
E-mail address: chowzhoumd@gmail.com (Q. Zhou).

http://dx.doi.org/10.1016/j.radcr.2016.04.012

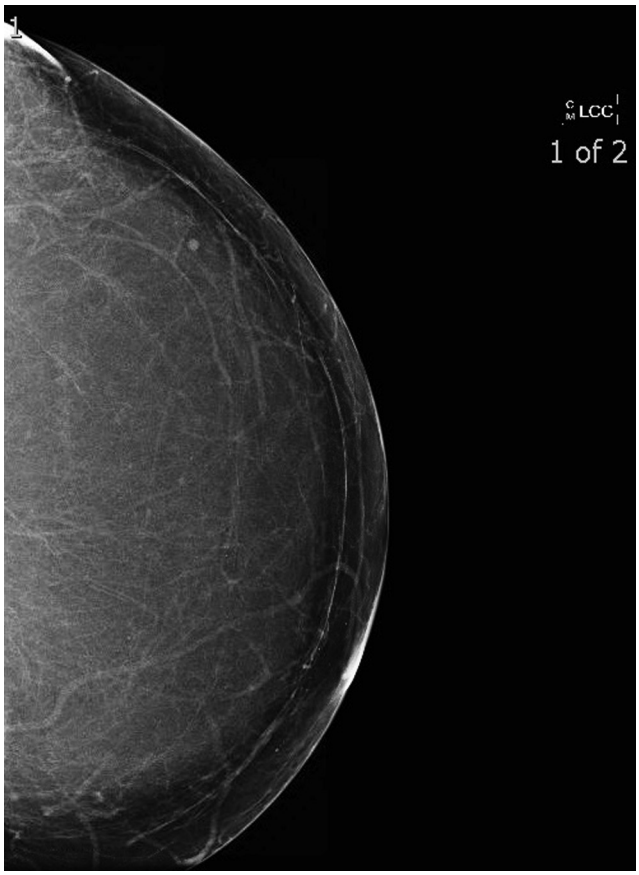


**Fig. 1 – 7/3/2014: Mammogram demonstrates no evidence of malignancy.**

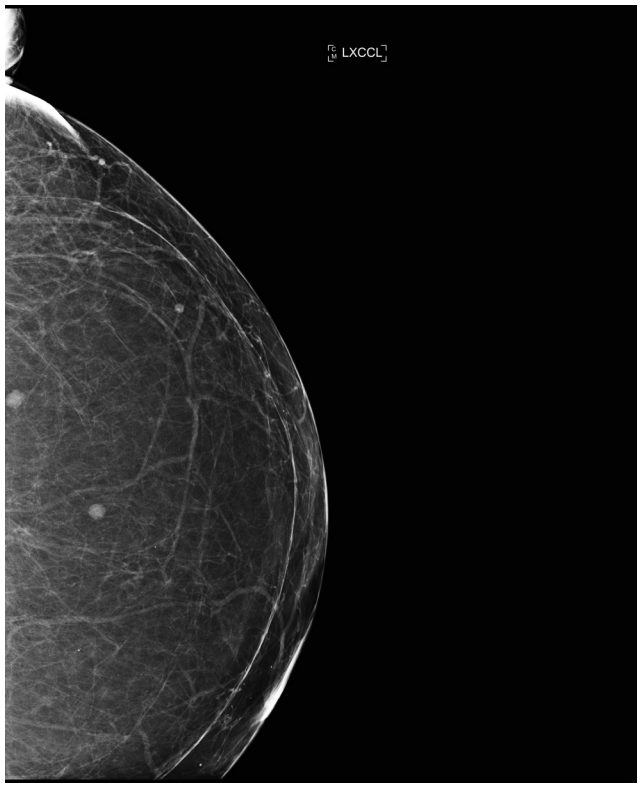


**Fig. 2 – Mammogram 7/9/2015: 2 new nodules within the upper left breast, one at the 12:00 position and the other at the 1 to 2:00 position posteriorly.**

systemic lymphadenopathy. Because of the patient's history of breast carcinoma and isolated locus of disease, it was ultimately determined by the patient's multidisciplinary team to perform a curative resection of the involved site.

## Discussion

Sinus histiocytosis and massive lymphadenopathy also known as RDD is characterized by an uncontrolled proliferation of the non-Langerhans sinus histiocyte [1]. Pathologically, these histiocytes harbor what are presumably engulfed, intact lymphocytes lending the appearance of "cell within a cell" or emperipolesis [2]. Identification of cell surface S-100 and CD68 receptors and the lack of CD1a expression are the cardinal histologic characteristics of this cell type [1]. This rare disease demonstrates predominance in young adults; however, adult cases have also been documented. Most patients present with profound cervical lymphadenopathy in addition to headaches and seizures if the intracranial space is involved. The most commonly reported extranodal sites include the skin, central nervous system, upper respiratory tract, orbit and eyelid, and gastrointestinal tract [3,4]. The exact inciting factor is unknown, but immune system dyscrasias and viral infections, specifically herpesvirus 6, have been implicated [5].

Infiltration into breast tissue by RDD is a very rare occurrence [6,7]. Moreover, disease isolated solely to the breast is rarer still, as RDD commonly presents with lymphadenopathy and involves multiple organs at the time of diagnosis [8]. Although RDD tends to present most commonly in young males (average age 39 years) involving multiple organs, most of the cases isolated to the breast occur in female patients older than 50 years [9]. This is especially significant because women after 50 years are screened regularly because of increased risk of breast carcinoma, and initial radiographic and clinical presentation of RDD can masquerade as lesions running the gamut of benign cysts and fibroadenomas to carcinoma [10,11]. Differential considerations of mass-like, histiocytic invasion into breast tissue should always include RDD among other entities such as Langerhans' cell histiocytosis, Erdheim-Chester disease, diabetic mastopathy, and granulomatous lobular mastitis [12,13].

In reference to the patient presented in this case, the discovery of 2 new nodular lesions in the left breast in the setting of right breast cancer status after mastectomy were cogent indications to proceed with stereotactic biopsy. RDD can have a high rate of recurrence which could potentially lead to repeated biopsies and excisions in the breast tissue [14]. Notably, incidental discovery of RDD in the breast parenchyma necessitates establishing the full extent of disease elsewhere in the body with full body CT and abdominal ultrasound [15]. Positron emission