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CASE REPORT

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Bilateral breast Rosai-Dorfman disease screen detected by mammography

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Abstract

Rosai-Dorfman disease (RDD) is a proliferative disorder of histiocytes typically found in nodal sites and commonly observed in females. Patients often present with systemic symptoms such as fever, lymphadenopathy, and weight loss. However, extra-nodal disease has been identified in locations including the skin and subcutaneous tissue. We present a case of a 59-year-old female presenting with abnormal bilateral findings on screening mammography, who was found to have a rare presentation of Rosai-Dorfman disease.

KEYWORDS

breast imaging, breast ultrasound, histiocytosis, mammography, Rosai-Dorfman disease

1 | INTRODUCTION

Rosai-Dorfman disease (RDD) or sinus histiocytosis with massive lymphadenopathy is a proliferative disorder of histiocytes of uncertain pathogenesis typically found in nodal and, less commonly, in extra-nodal sites such as the skin, subcutaneous tissue, bone, central nervous system, breast, thyroid, and mesentery.[1]

The disease has a female predilection with presenting symptoms including fever, lymphadenopathy, elevated erythrocyte sedimentation rate, leukocytosis, weight loss, polyclonal hypergammaglobulinemia, and anemia.[1] The etiology is still unknown, but theories suggest an immune regulation disorder or infections such as Herpes virus, Epstein–Barr virus, Cytomegalovirus, Varicella Zoster virus, or HIV.[2]

The histological appearance is similar regardless of the site of occurrence with the extra-nodal lesions demonstrating more fibrosis.[3] Histologic findings include a benign proliferation of histiocytes with abundant cytoplasm and vesicular nuclei, and histiocytes containing engulfed intact lymphocytes in a background of mature lymphocytes and plasma cells.[4] Characteristic histologic findings are histiocytes that stain for S-100 protein, OCT2, cyclin D1, and CD163.[5,6]

The overall prognosis of RDD is favorable, however, patients with solid organ involvement may require surgical intervention and carry a worse prognosis.[2,5] Treatments include corticosteroids, surgery, radiation therapy, chemotherapy, and immunomodulatory therapy. Some patients opt for an expectative approach and monitoring.[2] Of note, RDD, or sinus histiocytosis with massive

lymphadenopathy, can affect the nasal septum and present as nasal congestion or obstruction secondary to a septal or paraseptal mass. These cases are typically managed through endoscopic surgical resection. Further, benign sinonasal tumors tend to display a higher quality of life (QOL) pre- and postoperatively with the use of minimally invasive endoscopic resection, whereas the use of radiation and/or chemotherapy is associated with less favorable outcomes.[7]

2 | CASE PRESENTATION

A 59-year-old female was recalled from screening mammogram for abnormal findings in both breasts. Her medical history was pertinent for Hypertension and Type II Diabetes. Her surgical history was pertinent for a prior benign right breast biopsy, and a prior benign left excisional biopsy. She had no known personal history of hematologic disorders.

Her most recent prior mammogram was 3 years prior and was negative (BI-RADS 1). Routine screening mammography showed a round mass in the upper outer quadrant of the right breast and a focal asymmetry in the medial left breast. These findings were identified on medial lateral oblique (Figure 1) and cranio-caudal (Figure 2) views.

The patient underwent targeted bilateral breast ultrasounds for each finding. Ultrasound images demonstrated an irregular solid mildly vascular mass with angular margins measuring up to 1.4 cm in the right breast at 10 o'clock (Figure 3A). This finding corresponded to the mass seen on mammography. In the left breast, two irregular hypoechoic and hyperechoic solid masses were visualized at 10 o'clock with the largest measuring 1.1 cm (Figure 3B).

These masses were considered suspicious, and an ultrasound-guided biopsy of the right breast mass and the larger of the two left breast masses was recommended and ultimately performed. Pathology from both masses revealed atypical histiocytic proliferation. Immunohistochemical staining demonstrated histiocytes positive for S-100, OCT2, cyclin D1, and CD163 (Figure 4). These features are consistent with Rosai-Dorfman Disease and the diagnosis was made. The patient was followed with close observation by Hematology and Oncology and is doing well without clear evidence of critical organ involvement or inflammatory symptoms.

3 | DISCUSSION

Rosai-Dorfman disease limited to the breasts is extremely rare with only 17 cases reported in English literature.[8] RDD can mimic breast cancer clinically and on imaging studies, thus leading to surgical excision in most cases. The cases that have been reported in the literature have radiographically presented as breast lesions suspicious for malignancy.[9] Patients with RDD confined to the breast may present with a palpable mass, pain/tenderness, or an abnormal screening mammogram. Mammogram findings may show a single or multiple ill-defined mass(es) without calcifications. Ultrasound typically demonstrates a hypoechoic mass with indistinct or angulated margins and increased vascularity on color Doppler imaging.[10] The presentation is concerning given its mammographic features of a suspicious abnormality (BI-RADS 4), such as invasive carcinoma.[8] Definitive diagnosis is established only through tissue sampling.[5] The disease usually spontaneously resolves and follows a benign course, however, in few cases it may progress

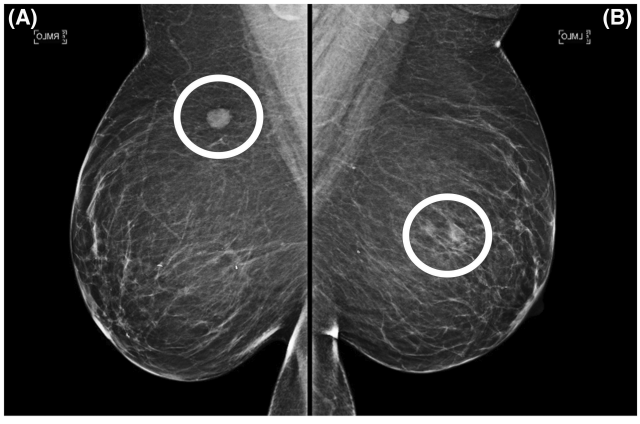

FIGURE 1 (A) Right breast medial lateral oblique view showing a round mass in upper breast not previously seen on prior studies. (B) Left breast medial lateral oblique view showing focal asymmetry not previously seen on prior studies.

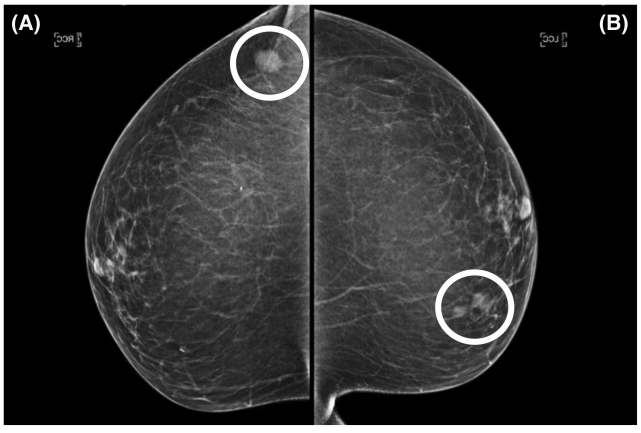

FIGURE 2 (A) Right breast cranio-caudal view showing mass in outer breast not evident on prior studies. (B) Left breast cranio-caudal view showing focal asymmetry in inner breast not evident on prior studies.

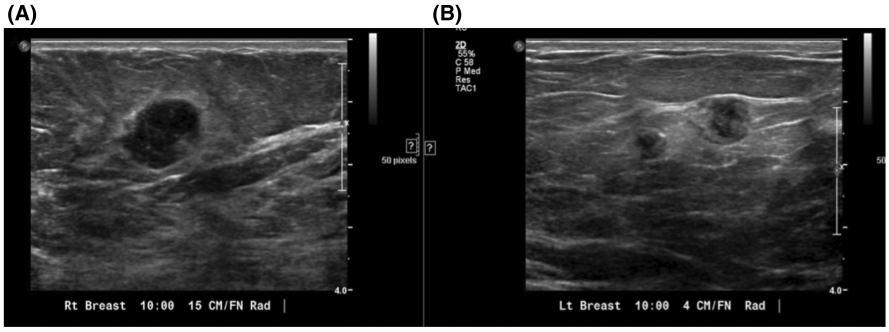

FIGURE 3 (A) Right breast ultrasound at 10 o'clock 15 cm from the nipple showing an irregular solid mass with angular margins. (B) Left breast ultrasound at 10 o'clock 15 cm from the nipple demonstrating two irregular hypo- and hyperechoic solid masses.

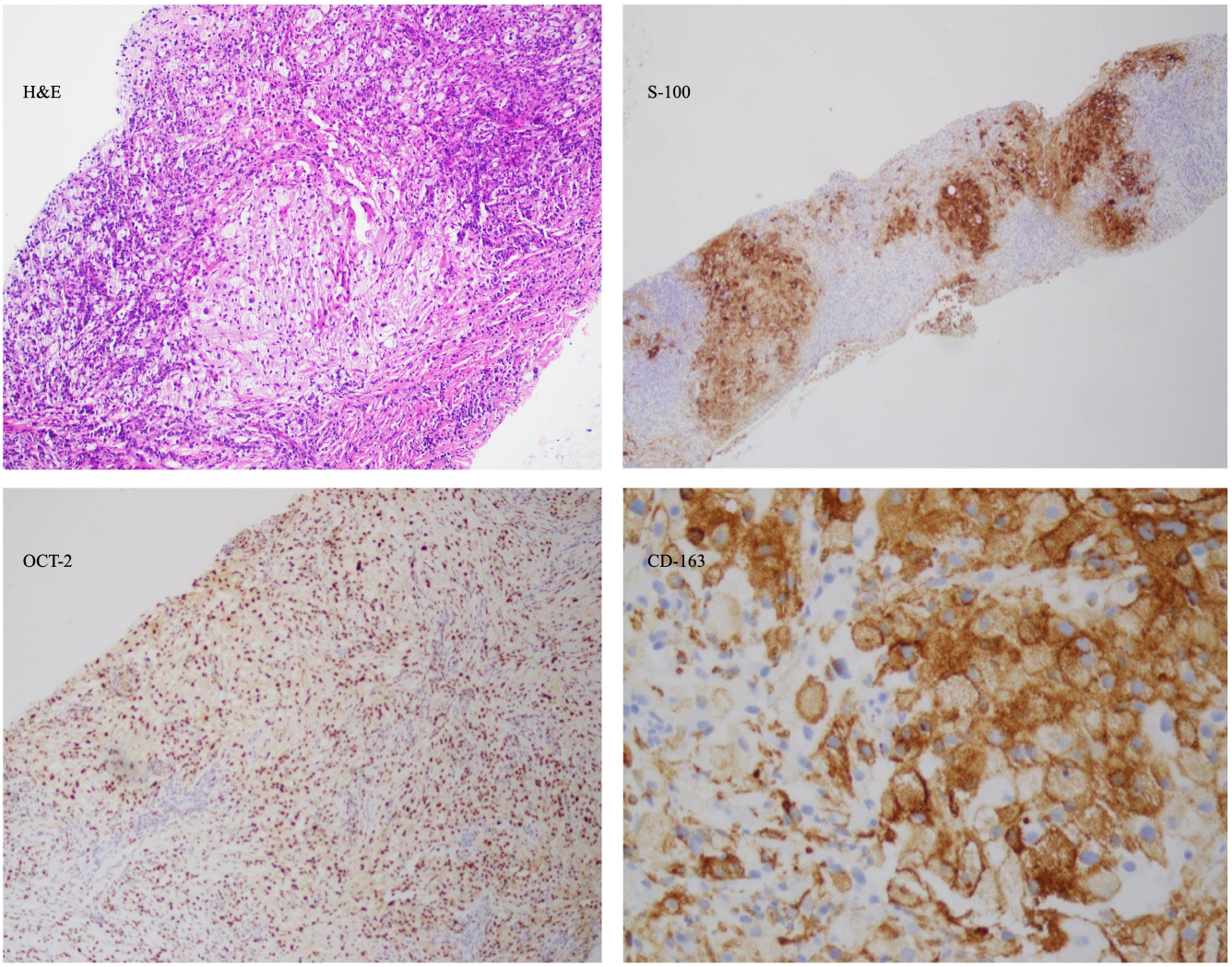

FIGURE 4 Breast mass pathology demonstrating atypical histiocytes on H&E staining, with positive staining for S-100, OCT-2, and CD-163.

with solid organ involvement. Close surveillance is thus recommended.

AUTHOR CONTRIBUTIONS

Christina Sumner: Conceptualization; data curation; formal analysis; project administration; writing – original draft. **Karma Salem:** Data curation; formal analysis. **Luma Abunimer:** Writing – review and editing. **Abdulwahab Ewaz:** Data curation; formal analysis. **Linsheng Zhang:** Data curation; formal analysis. **Ashley Monsrud:** Data curation; formal analysis. **Nabil Calisi:** Conceptualization; data curation; formal analysis; project administration.

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CONFLICT OF INTEREST STATEMENT

The authors declare that no conflict of interest exists.

DATA AVAILABILITY STATEMENT

Data sharing not applicable to this article as no datasets were generated or analysed during the current study

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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