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The Halo Sign on Ultrasonography of Breast Nodules May Be Benign: A Case Report of Rosai-Dorfman Disease Confined to Breast

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Abstract

Roussel-Dorfman disease (RDD) is a histiocytosis that usually presents in young adults or children and presents with bilateral cervical lymphadenopathy, but extranodal involvement is also rare, especially in the breast. A 41-year-old woman presented with breast tenderness and palpable lumps in both breasts. Ultrasonography showed: multiple nodules in the bilateral breasts with unclear borders, irregular shapes, hyperechoic halos (the halo sign) around the edges, and dilated ducts under the nipples. Our case showed bilateral breast disease with the suspicion of breast cancer on ultrasonography. However, pathological analysis showed large histiocytic proliferation and interstitial fibrosis, with positive S-100 and CD68 immunoreactivity. RDD of the breast is a rare form of histiocytosis, the sonographic appearance can be similar to breast cancer. Thus, a multidisciplinary team decision is important.

Keywords: *Rosai-Dorfman disease, histiocytosis, halo sign, ultrasonography.*

Introduction

Roussel-Dorfman disease (RDD) is a histiocytosis that commonly affects the cervical lymph nodes in children and young adults. However, extranodal involvement is not uncommon and often complicates diagnosis and treatment. In 1969, Rosai and Dorfman correctly identified the critical role of histiocytes in the pathogenesis of the disease, which typically manifests as bilateral painless cervical lymphadenopathy [1]. The most common extranodal localizations reported in the literature are the nasal cavity, skin, orbit, bones, and central nervous system, but also include skeletal muscle, subcutaneous tissue, heart, or thyroid. Breast localization is rare and often leads to difficult differential diagnosis with breast malignancy [2]. We report the clinicopathological features of extranodal RDD, presenting as a palpable breast mass with sonographic similarities to breast cancer.

Case presentation

A 41-year-old woman presented to our outpatient clinic complaining of painful, palpable lumps in both breasts over the past week. She has no history of breast disease or chronic diseases such as hypertension or diabetes. Blood laboratory tests revealed no obvious abnormalities. Ultrasonography showed: the ducts under the bilateral breast nipples were dilated, and multiple hypoechoic nodules were seen in both breasts. One of the left breasts at 8 o'clock was about 0.7X0.4cm in size, with clear borders and regular shape; 10 o'clock position about 2.0cm away from the nipple was about 0.9X0.6cm in size. The boundary was unclear and the shape was irregular. A hyperechoic halo (the halo sign) could be seen around the nodules. Color Doppler Flow image (CDFI): punctate blood flow signals were seen (**Figure1)**.

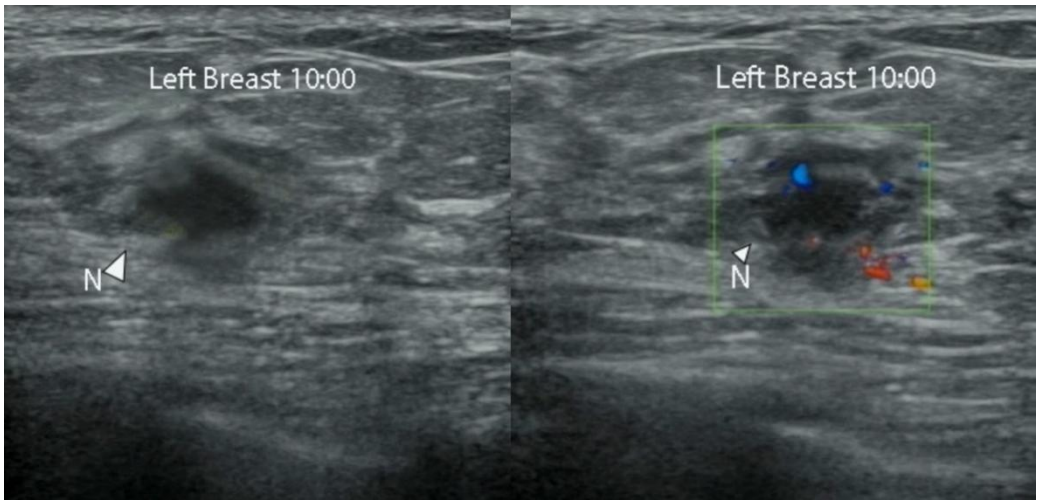


Figure 1: Ultrasonography: Hypoechoic nodule in the left breast at 10 o'clock, unclear boundary, the halo sign visible on the periphery, and punctate blood flow signals seen on CDFI. N: nodule

The size of the right breast at 6 o'clock is about 1.1X0.6cm, the border was clear, the shape was irregular, the halo sign could be seen around the nodule, CDFI: punctate blood flow signals were seen (**Figure2).**

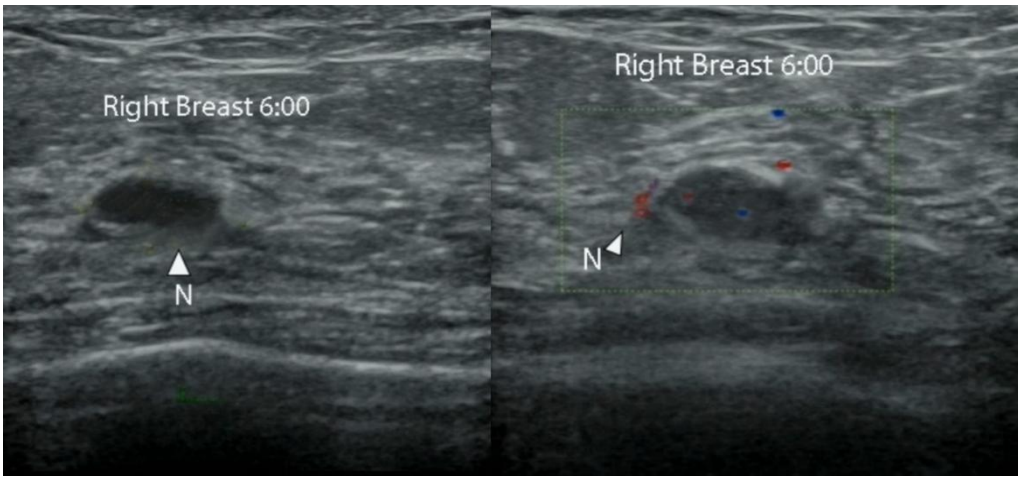
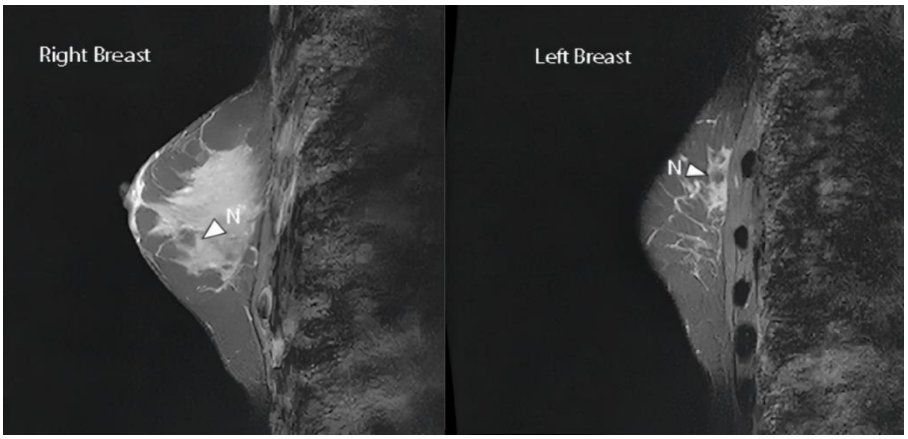


Figure 2: Ultrasonography: Hypoechoic nodule in the left breast at 6 o'clock, unclear boundary, the halo sign visible on the periphery, and punctate blood flow signals seen on CDFI.

No abnormal structural lymph nodes were detected in the bilateral axillary. The nodules at 10 o'clock on the left breast and 6 o'clock on the right breast were considered to be BI-RADS-4a, and the remaining nodules were classified as BI-RADS-3. Magnetic resonance imaging (MRI) showed: local duct expansion in both breasts, patchy asymmetric short T1 and long T2 signals with unclear boundaries in the upper inner quadrant of the left breast, enhanced scan showing non-mass-like obvious enhancement, and nodules seen directly behind the right breast, which appeared Long T1 and long T2 signal, the edge of the nodule showed shallow lobulation and spiky changes, and the enhanced scan showed heterogeneous high enhancement (**Figure 3& Figure 4).**



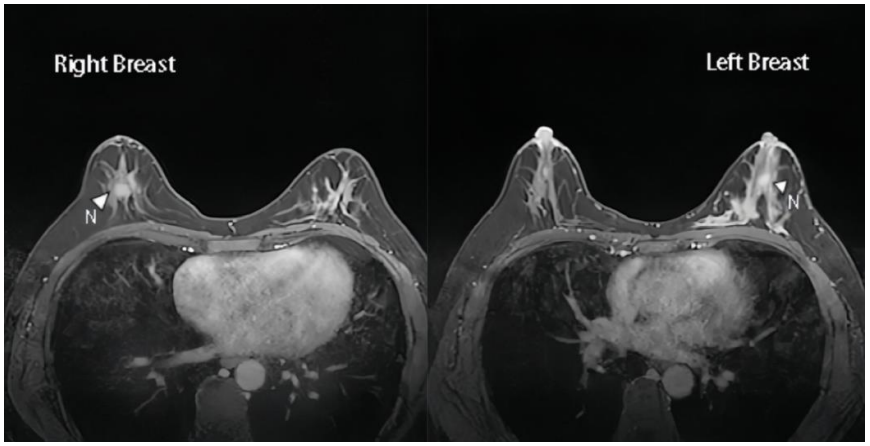


Figure 3 & Figure 4: MRI: Flake-like abnormal signals were seen in the upper inner quadrant of the left breast with unclear boundaries, and the enhanced scan showed obvious enhancement. Nodular abnormal signals were seen directly behind the right breast, with shallow lobulation and burr-like changes at the edge. The enhanced scan showed heterogeneous and obvious enhancement.

MRI also classified the nodules into BI-RADS-4a. Mammography showed: asymmetric patchy shadow in the upper quadrant of the left breast, but no obvious abnormality in the right breast (**Figure 5).**

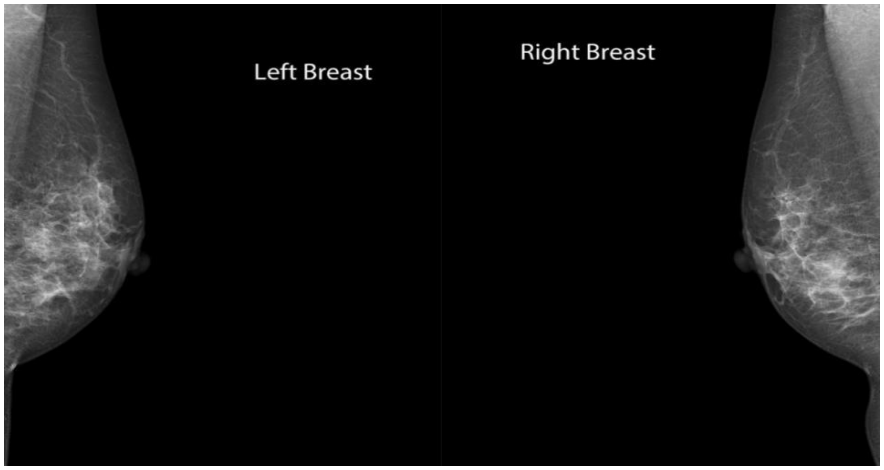


Figure 5: Mammography: no obvious signs of malignancy.

Radiologists classify it as BI-RADS-3. The surgeon considered the possibility of malignancy in the breast nodules and therefore performed surgical resection of the breast nodules. Pathology exam revealed histiocyte aggregates, emperipolesis, histiocytes diffuse positive for S100 protein, CD1a, focally expressing CD68, negative for CD20 and CD3 (**Figure 6).** Pathological results were consistent with a diagnosis of Rosai-Dorfman disease of the breast. Unfortunately, we don't have long-term follow-up information on the patient.

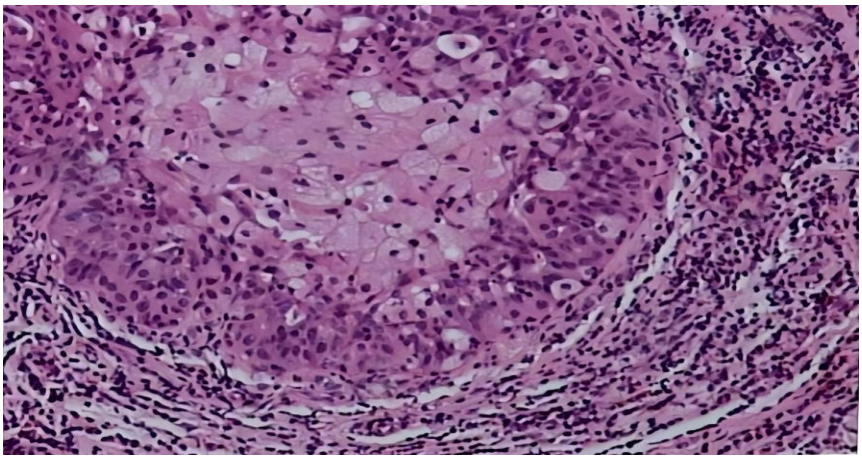


Figure 6: Pathology: histiocyte aggregates, emperipolesis, histiocytes diffuse positive for S100 protein, CD1a, focally expressing CD68, negative for CD20 and CD3.

Discussion

Rosai–Dorfman disease (RDD) is defined as a self-limiting proliferative disorder of non-Langerhans histiocytes, usually occurring in isolated lymph nodes, usually in young male patients, without racial predilection [3]. The cause of RDD is unknown, but some reports suggest that viruses such as human herpesvirus, parvovirus B19, and Epstein-Barr virus may be involved [4]. Common clinical symptoms include fever and weight loss, and elevated white blood cells and erythrocyte sedimentation rate. In our case, laboratory blood test results were within normal limits. Although RDD is usually a self-limiting benign disease, it can manifest with multi-organ involvement and some patients have a poor prognosis [5]. It has been reported in the literature that patients with RDD with systemic multi-organ involvement should undergo whole-body scans, including computed tomography (CT) of the chest and pelvis or positron emission tomography-computed tomography (PET-CT), to evaluate for other RDD with extranodal involvement [6].

Since RDD is a rare disease, it may be mistakenly considered a malignant disease on clinical examination, imaging, and even pathology [6]. Most cases of lesions confined to the breast occur in women over 50 years old. In particular, involvement of axillary lymph nodes can cause additional confusion for clinicians [7, 8]. The imaging manifestations of RDD are diverse, which can range from those of a simple cyst to those of a breast carcinoma [9]. RDD may be mistaken for a malignant disease on clinical examination, imaging and even pathology. In our case, the patient was younger than reported in the literature. Ultrasonography revealed the halo sign around the breast nodule, which is highly suspicious of malignancy in the BI-RADS scoring system [10]. At the same time, MRI showed the spiculation sign around the nodule, which prompted the surgeon to choose surgical resection, although the mammography showed no signs of malignancy. Before surgical resection, core needle biopsy may be a more appropriate method. Shin et al reported that clear halo of cytoplasm surrounding the lymphocytes may be the pathological basis for the halo sign of nodules in ultrasonography.

The differential diagnosis for RDD includes Langerhans' cell histiocytosis, granulomatous lobular mastitis, IgG4-sclerosing mastitis, breast lymphoma with plasmocytic differentiation, Erdheim-Chester disease, and diabetes mellitus mastopathy [11]. In most cases, it is not necessary to treat RDD confined to the breast. However, due to lack of data in the literature, many patients undergo surgical resection or tumor resection for diagnostic and therapeutic purposes [12].

Result

Due to both ultrasonography and MRI suggesting the possibility of malignancy in the breast nodules, the patient underwent surgical resection of the nodules. There was excessive medical treatment here, and core needle biopsy should be a more appropriate method. Unfortunately, the patient did not undergo follow-up and reexamination at our hospital, and we are unable to determine whether it has recurred.

Conclusion

Extranodal manifestations of RDD involving the breast are rare benign conditions that are difficult to distinguish clinically and radiographically from breast malignancies. It is important to recognize the unique pathological features of RDD to avoid unnecessary invasive procedures in patients. Thus, a multidisciplinary team decision is important.

Conflict of Interest

The authors declare that they do not have anything to disclose regarding funding or conflict of interest with respect to this manuscript.

Ethical statements

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1964 and later versions. Informed consent was obtained from the patient for publication of this case report.

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Author contributions:

Conceptualization: Xiangyu Chen.
Data curation: Ling Wang, Xiangyu Chen.
Writing – original draft: Ling Wang.
Writing – review & editing: Xiangyu Chen.

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