Case Report

Eur J Breast Health 2024; 20(3): 228-230
DOI: 10.4274/ejbh.galenos.2024.2024-1-9

A Rare Case of Concurrent Lupus Mastitis and Sarcoidosis in a 62-Year-Old Female

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ABSTRACT

Systemic lupus erythematosus (SLE) and sarcoidosis are two of the most well-recognized, chronically diagnosed conditions in the United States, with a plethora of known multisystem manifestations. With regard to breast pathology, lupus mastitis is a relatively uncommon manifestation of SLE, commonly involving both the mammary gland and subcutaneous soft tissues of the breast. Sarcoidosis in the breast is a similarly, exceedingly rare manifestation of this multi-system disorder, classically presenting with non-caseating granulomas. Both present with non-specific mammographic and sonographic features. We present a 62-year-old female with known diagnosis of discoid lupus and Graves' disease who presented initially with an abnormal screening mammogram, ultimately undergoing mammographic work-up and subsequent biopsy demonstrating lupus mastitis, including vasculitis, panniculitis, and fibrosis with chronic inflammation. The patient was also found to have small non-caseating granulomas, some in a perivascular distribution, classically seen in sarcoidosis. Given the rarity of both manifestations, our case explores the coexistence of these autoimmune processes and this atypical presentation.

Keywords: Lupus mastitis; systemic lupus erythematosus; breast sarcoidosis; granulomatous disease; mammogram; breast ultrasound; ultrasound-guided core needle biopsy

Cite this article as: Fofana CC, Del Toro C, Alvarez-Lesmes J, Pluguez-Turull CW. A Rare Case of Concurrent Lupus Mastitis and Sarcoidosis in a 62-Year-Old Female.

Key Points

- Multisystem inflammatory processes occurring in the breast, albeit rare, can occur in tandem and present confounding diagnosis and clinical presentation, imaging, and pathologic diagnoses.
- Chronic inflammatory processes in the breast can present suspicious mammographic and ultrasound findings, mimicking malignancy. Further research can lend into characteristic imaging findings.
- Established treatment algorithms for chronic inflammatory processes such as sarcoid and lupus are not clearly defined. Further research can explore optimizing appropriate strategies for relief.

Introduction

Sarcoidosis is a systemic inflammatory disease of unknown etiology classically characterized by the formation of multiorgan granulomatous lesions. Existing literature has cited an immune mediated secondary response to an antigenic trigger as a potential etiology (1). In the United States, the disease is most prevalent in African Americans, with prevalence rates of up to 10 times that of white Americans (2). Extensive variability is demonstrated as far as disease prognosis, manifestations, and progression of disease. The mainstay of treatment is corticosteroids, although in severe cases immunosuppressive agents, such as methotrexate and cyclophosphamide, are used. Sarcoidosis of the breast is rare and only few cases are reported in the literature. Mammographic features are variable and may mimic both benign and malignant etiologies. The breast is involved in less than 1% of cases and can either be a primary or secondary site of presentation (3). Breast sarcoidosis can mimic carcinoma on mammographic and clinical exam, as seen in the presented case (4).

Lupus is characterized as an autoimmune disease in which a misdirected immune response generates a multisystem inflammatory response against the native tissues. Similar to sarcoidosis, the disease prognosis and manifestations are extremely variable, ranging from mild to severe. Lupus mastitis is an uncommon presentation of systemic lupus erythematosus (SLE), characterized by inflammation of the adipose fat. Lupus mastitis can present as single or multiple subcutaneous or deep breast masses, often clinically mimicking malignancy (5). The constellation of concurrent lupus and sarcoidosis in the breast has not been previously reported. Given the suspected immune mediated etiology, the intersection of concurrent pathology in our case may yield

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Received: 25.01.2024
Accepted: 14.04.2024
Available Online Date: 01.07.2024

as a branch point to observe how systemic processes can present and mimic each other.

Case Presentation

A 63-year-old female with past medical history of discoid lupus and Graves' disease, with no known diagnosis of sarcoidosis at presentation, was found to have an indeterminate asymmetry in the right breast at 12 o'clock anterior depth (Figure 1), persisting on spot compression views (Figure 2). Clinically, no lesions were palpated, and the patient did not report any pain in the right breast. In addition, views with ultrasound demonstrated a 4.4 x 1.9 x 3.7 cm indistinct area of mixed echogenicity lacking significant flow on power Doppler in the right breast, corresponding to the partially persistent asymmetry on mammogram (Figure 3). A palpable second area of concern in the left breast prompted ultrasound, which revealed a 4.7 x 1.6 x 3.9 cm area of irregular, heterogenous, non-mass lesions lacking significant flow on power Doppler corresponding to a clinically reported palpable abnormality in the left breast (Figure 4). At the time, with a BI-RADS 4B classification for both areas in the breast, the decision was made to further evaluate the lesions via ultrasound-guided core needle biopsy. Pathology revealed scattered small non-caseating granulomas with giant cells on a background of chronic inflammation in the left breast with concurrent focal vasculitis and panniculitis. The right breast similarly demonstrated small non-caseating granulomas with giant cells on a background of chronic inflammation, panniculitis and more prominent focal vasculitis. These findings were seen in the context of fibrosis and chronic inflammation (Figures 5 and 6) and led to a diagnosis of lupus mastitis with recommendations for clinical correlation and consideration of sarcoidosis. The patient was subsequently referred to a rheumatology clinic and was found to have nodular lesions on the nose, which can be seen in sarcoidosis, however, with normal angiotensin converting enzyme level and serum anti-nuclear antibodies, extracatable nuclear antigen antibodies and double-stranded DNA antibody titers. Additional laboratory results showed leukopenia and neutropenia both of which were chronic and had been stable. Chest computed tomography imaging demonstrated

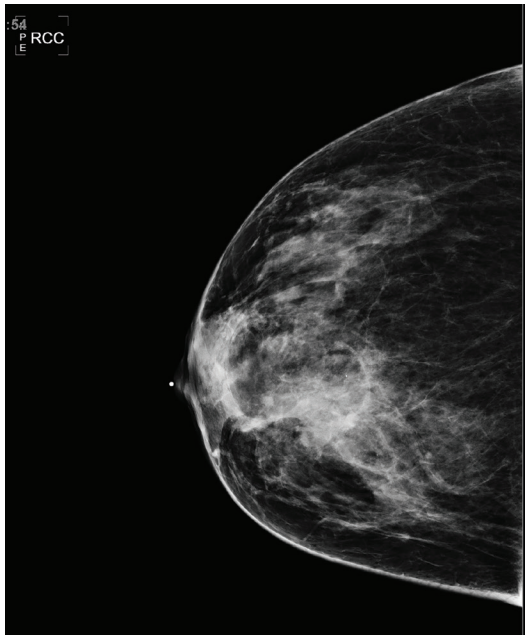

Figure 1. Initially reported indeterminate asymmetry in the right breast at 12 o'clock anterior depth

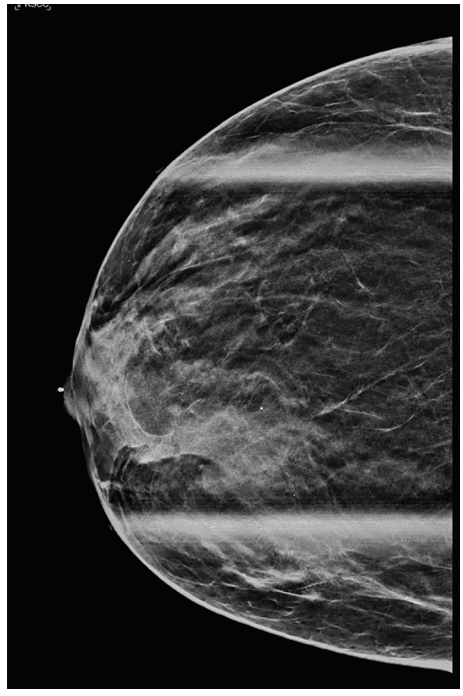

Figure 2. The previously seen focal asymmetry in the right breast anterior third at the 12:00 axis partially persists on additional views

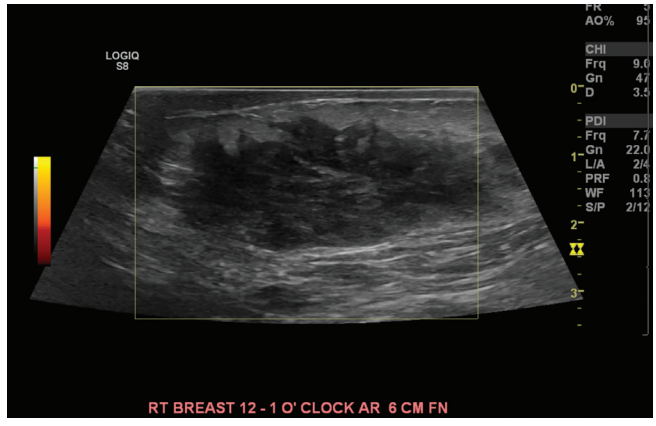

Figure 3. A 4.4 x 1.9 x 3.7 cm indistinct area of mixed echogenicity in the right breast at 1:00, 6 cm from the nipple, lacking significant flow on power doppler in the right breast, corresponding to partially persistent asymmetry on mammogram

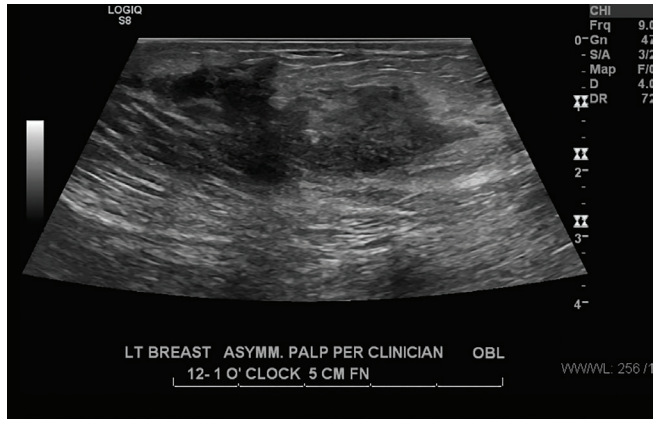

Figure 4. 4.7 x 1.6 x 3.9 cm irregular area of heterogeneity at 12:00-1:00, 5 cm from the nipple, lacking significant flow on power doppler corresponding to a palpable abnormality in the left breast

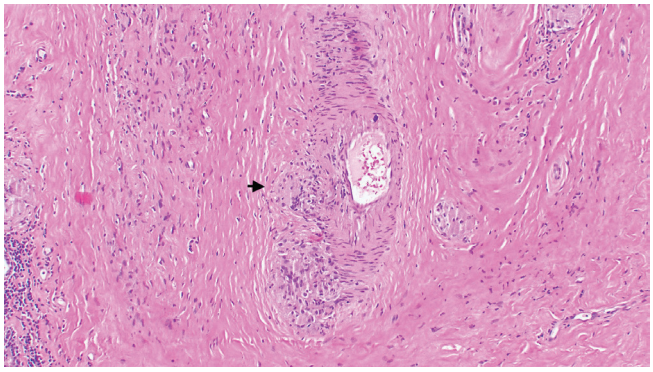

Figure 5. Focus of vasculitis with adjacent granulomatous inflammation (arrow), right breast. 100x magnification

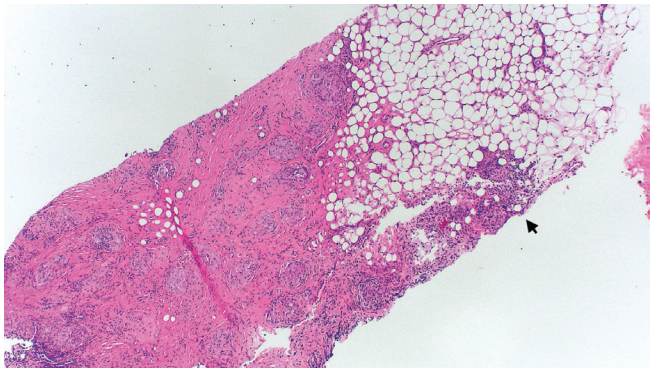

Figure 6. Benign breast parenchyma with non-caseating granulomas and giant cells (arrow) and background chronic inflammation, right breast. 40x magnification

mediastinal lymphadenopathy and multiple pulmonary nodules and fibrotic changes, which can be seen in sarcoidosis. Subsequent positron emission tomography exam demonstrated active uptake of these lesions, suggestive of active disease. These imaging findings along with the biopsy results provided enough evidence to support the diagnosis of sarcoidosis and thus a biopsy of the nose was not recommended by dermatology for cosmetic reasons. The concurrent diagnosis of sarcoidosis of the breast was made.

Outcome and Follow-up

Patient was subsequently started on 10 mg of methotrexate and folic acid and continues long-term follow-up in rheumatology clinic.

Discussion and Conclusion

The multisystem manifestations of SLE and sarcoidosis have been well-documented in the literature. However, in both systemic processes, breast manifestations are extremely rare. In addition, the concurrent presentation of both lupus and sarcoidosis has rarely been reported previously (6). Variable mammographic and sonographic imaging patterns have been seen in both processes, further complicating diagnosis and timely recognition (4). This case is atypical in that the mammographic findings ultimately yielded an unsuspected diagnosis of sarcoidosis in the setting of an asymptomatic presentation, and initiation of long-term therapy. Given the expanding research regarding risk factors for autoimmune pathology in chronic illness, our case serves as an exploration into the manifestations of autoimmune processes. The delineating factor in this case was the histopathological diagnosis, as the mammographic and sonographic findings were nonspecific. Initial sonographic findings were concerning for potential malignancy, further emphasizing the variable appearance of systemic autoimmune processes within the breast. Further research may serve to analyze risk factors for breast manifestations of these processes and treatment algorithms for appropriate management in the setting of chronic disease. Given the low incidence of breast manifestations, obtaining enough cases may serve as a research limitation. Further research may serve to analyze sonographic, mammographic and possibly histopathological features of autoimmune manifestations within the breast to assess if recurrent characteristics are visualized.

Informed Consent: Written informed consent was obtained from the patient.

Authorship Contributions

Surgical and Medical Practices: C.C.F., J.A-L., C.W.P-T.; Concept: C.C.F., C.D.T., J.A-L., C.W.P-T.; Design: C.C.F., C.D.T., J.A-L., C.W.P-T.; Data Collection and/or Processing: C.C.F., J.A-L., C.W.P-T.; Analysis or Interpretation: C.C.F., C.D.T., J.A-L., C.W.P-T.; Literature Search: C.C.F., C.D.T., J.A-L., C.W.P-T.; Writing: C.C.F., C.D.T., J.A-L., C.W.P-T.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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