Case Report

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Lupus Mastitis Manifesting as Extensive Calcification in the Retromammary Area: A Case Report

유선후층의 광범위한 석회화로 나타난 루푸스 유방염: 증례 보고

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Lupus mastitis is a presentation of lupus panniculitis that involves subcutaneous fat in patients with systemic lupus erythematosus (SLE). Moreover, lupus mastitis is a rare condition that typically presents as a palpable breast mass. Here, we report the case of a 29-year-old woman with a prior diagnosis of SLE who presented with palpable lumps in both breasts. Imaging studies were performed, and mammography revealed extensive and bizarre calcifications, mainly in the retromammary area. US revealed large irregular calcifications with posterior shadowing, and breast MRI images exhibited diffuse irregular persistent rim enhancement in the retromammary area. An US-guided biopsy was performed to differentiate the condition from breast cancer and confirmed the diagnosis of fat necrosis, consistent with the pathological characteristics of lupus mastitis. Herein, we present a case of lupus mastitis, an unusual clinical manifestation of SLE, and discuss the clinical, pathological, and imaging findings supporting the diagnosis and the differential diagnosis.

Index terms Lupus Mastitis; Fat Necrosis; Retromammary Area; Mammography; Magnetic Resonance Imaging

INTRODUCTION

Breast tissue can occasionally be affected by extramammary systemic diseases such as collagen vascular diseases (1). Lupus mastitis is a rare clinical manifestation

of systemic lupus erythematosus (SLE) involving the breast. Lupus mastitis is a type of lupus panniculitis, which is inflammation of the deep subcutaneous fat layer due to SLE, occurring in the breast. Patients often report palpable masses in the breast and this condition tends to have a relatively chronic course (2).

The inflammatory response in this condition, which leads to fat necrosis in the breasts, often presents imaging findings that are atypical and may sometimes require differentiation from malignancy. Herein, we report a case of lupus mastitis presenting as a breast mass with unique, bizarre, and sheet-like extensive calcifications in the retromammary fat area on mammography.

CASE REPORT

A 29-year-old woman visited our breast surgery outpatient clinic with palpable breast masses. About a year ago, she had been diagnosed with SLE based on recurrent facial rashes, arthritis, leukopenia, proteinuria, and positive antinuclear antibody. The patient was treated with a low dose of glucocorticoid and immunosuppressive medications. Several months prior, she discovered palpable lumps in both breasts. During physical examination, multiple firm lumps were palpable in both breasts. However, she had no history of receiving foreign material injections into the breast.

Mammography revealed bizarre, sheet-like, and extensive calcifications of a dystrophic nature in both breasts. Although these lesions were diffuse, they were mainly located in the retromammary fat area (Fig. 1A). US revealed heterogeneous echoic lesions with posterior acoustic shadowing that correlated with extensive dense calcifications (Fig. 1B). Owing to the limitations of whole-breast evaluation by posterior shadowing on US, MRI was performed. Contrast-enhanced T1 axial imaging revealed irregular tubular branching-shaped masses with persistent rim enhancement and an internal fat component (Fig. 1C). The lesions had retracted the pectoralis muscle. Diffusion-weighted imaging did not demonstrate definite diffusion restriction, and no abnormal findings were identified in either axillary region. These findings were consistent with extensive fat necrosis; however, completely excluding the possibility of malignancy was challenging. US-guided core needle biopsy revealed inflammatory cell infiltration with dysmorphic calcification and fat necrosis, indicating lupus panniculitis (Fig. 1D).

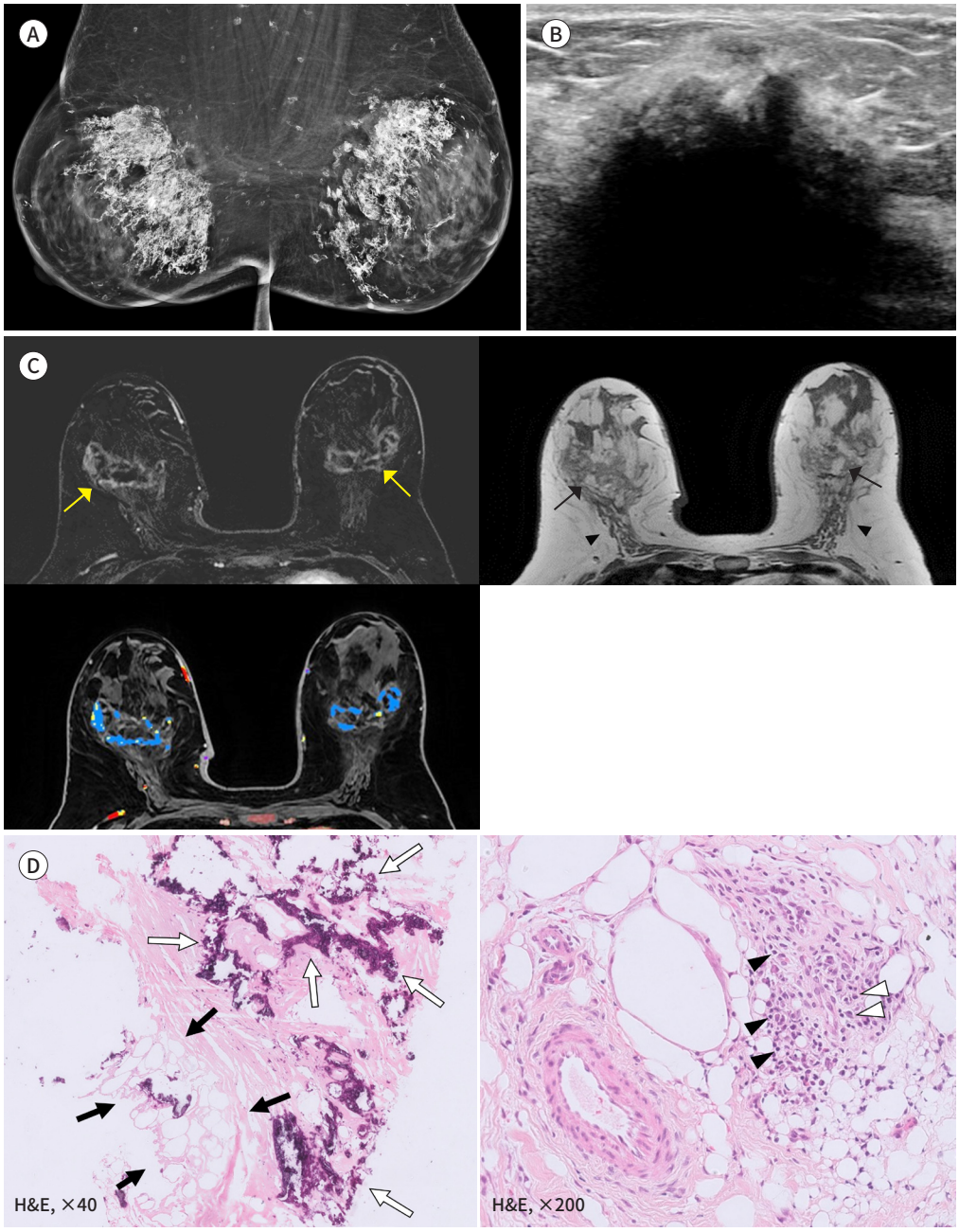
Based on the imaging findings, pathology results, and patient history, a diagnosis of lupus mastitis was made. Further surgical intervention for the breast lesions was deemed unnecessary. The patient decided to continue medication for her underlying disease and underwent outpatient follow-up.

This study was approved by the Institutional Review Board of the Kyungpook National University Chilgok Hospital (IRB No. 2023-10-013). Written informed consent was waived for the publication of this case report and accompanying images.

DISCUSSION

SLE is a representative autoimmune disorder that causes inflammation and damage to var-

Fig. 1. A 29-year-old woman diagnosed with lupus mastitis.
A. The mediolateral oblique view of mammography shows diffuse, symmetric, bizarre, and dystrophic calcifications in both breasts, especially in the retromammary area as well as the subcutaneous fat layer.
B. The US reveals heterogeneous echoic lesions in the right upper outer breast. Acoustic shadow resulting from dense calcifications is noted.
C. Contrast-enhanced T1-weighted MRI subtraction axial imaging (left upper) and computer-aided diagnosis image (left lower) reveal irregular tubular branching-shaped masses with rim enhancement (yellow arrows). Most of the contrast-enhanced areas exhibit persistent kinetics (blue color). Pre-contrast T1-weighted MRI axial imaging (right) shows areas of high signal intensity within some of the lesions, indicating the presence of internal fat components (black arrows). Retraction of the pectoralis muscles is also visible (arrowheads).
D. Histopathologic features demonstrate fat necrosis (black arrows) and bizarre calcifications (white arrows) (left). Perivascular infiltration of inflammatory cells including lymphocytes (black arrowheads) and plasma cells (white arrowheads) is also observed (right).
H&E = hematoxylin and eosin



ious tissues, including the joints, skin, kidneys, brain, and blood. Lupus panniculitis, also known as "lupus profundus," refers to the involvement of the subcutaneous fat layer and was first described by Kaposi (3). This is a rare clinical manifestation, occurring in approximately 2%–3% of all patients with SLE, and may potentially involve the arms, buttocks, head, neck, and thighs. Even more rarely, lupus panniculitis can manifest in the subcutaneous fat tissue of the breast, known as "lupus mastitis" (2).

To the best of our knowledge, the precise pathophysiology of lupus mastitis remains unclear. The prevailing hypothesis suggests that akin to its impact on other organs, SLE has an autoimmune-related etiology. Supporting evidence includes the detection of immune complexes in areas of panniculitis (2). In patients without skin lesions, vasculitis is presumed to be the underlying cause (3).

Clinically, lupus mastitis often presents as palpable masses with or without associated pain (4, 5). Superficial lesions may be accompanied by skin changes such as atrophy, ulceration, or reddish-to-purplish discoloration (6). In the present case, the patient presented with multiple masses in both breasts. However, no visible skin lesions were observed, likely because most lesions were located deep within the breast rather than superficially, particularly in the retromammary area.

Ill-defined asymmetries or irregular masses corresponding to palpable lumps may be observed on mammography (5, 7). Calcifications can vary depending on the stage of disease progression, initially appearing as fine linear branching or amorphous microcalcifications indicative of suspicious findings. Over time, the calcifications increase in size and become coarser, suggesting a dystrophic nature indicative of fat necrosis, as seen in the present case (6). Although she was diagnosed with SLE a year ago, the extensive calcifications in the lesions suggest that breast involvement may have occurred over an extended period. On US, the lesions present as heterogeneous echoic masses with or without acoustic shadowing owing to dense calcifications. On MRI, the presence of irregularly margined masses with rim enhancement raises the suspicion of malignancy and fat necrosis (6, 8). T1-weighted imaging without fat suppression helps identify internal fat components, which can aid in diagnosing fat necrosis.

In our case, one notable observation was the distribution of the lesions. Lupus mastitis most commonly presents as lesions in the subcutaneous fat layer, similar to other forms of lupus panniculitis. However, in this case, the majority of lesions appeared to be dominant in the retromammary fat layer (2). Although the exact cause remains unclear, the abundance of fat tissue in her retromammary area suggests why the lesions are more prominent in this area.

Breast cancer is the most important differential diagnosis for lupus mastitis. Caution is warranted, especially when a mass or asymmetry appears without calcifications, or is accompanied by suspicious microcalcifications that can be observed in the early stages of the disease. Confirmation of the fat component on MRI can offer significant clues for ruling out breast cancer. In our case, fat components exhibiting high signal intensity on T1-weighted images were identified within the enhanced rim. Furthermore, unlike breast cancer, lupus mastitis is known to demonstrate improvement in MRI findings with optimized medical treatment. The rim enhancement thins out, becomes discontinuous, and eventually disappears on serial MRI scans (6). Nevertheless, if cancer cannot be ruled out, a biopsy may be

considered. Core needle biopsy is preferred over surgical biopsy, as invasive procedures are known to exacerbate lupus mastitis (2, 4).

When lupus mastitis presents with extensive, symmetric, and dystrophic calcifications that favor a benign nature, excluding malignancy is relatively easy. Extensive fat necrosis can occur following whole-breast trauma, including falls or car crashes, but rarely exhibits symmetry. In several autoimmune diseases other than SLE, such as dermatomyositis and mixed connective tissue disease, extensive fat necrosis with calcifications in the breast have also been reported (1, 9). Symptoms related to SLE, including arthritis and skin rash, are known to precede or be concurrent with lupus mastitis in most cases, as in this patient, and are expected to assist in the differential diagnosis (5).

The most important and characteristic histopathological finding in lupus mastitis is hyaline fat necrosis. Involvement of fat lobules by mature lymphocytes, including plasma cells, is the predominant inflammatory feature. Additionally, inflammatory cells can be detected in blood vessels, with lymphocytic vasculitis commonly affecting medium and small vessels (2).

Antimalarial drugs prescribed for skin lesions and arthritis in SLE patients can be beneficial for lupus mastitis. Furthermore, glucocorticoids and various immunosuppressive agents are also viable treatment options. Given that invasive examinations and treatments such as surgical excision can potentially trigger flares, they are recommended to be avoided. However, mastectomy can be considered as a final resort for patients experiencing severe pain unresponsive to medication (2, 8).

In conclusion, differentiating lupus mastitis from malignancy is important as the condition requires specifically tailored medical treatment. When distinctive breast calcifications and masses are encountered, as in this case, diagnosing lupus mastitis can be challenging due to its exceptionally low incidence rate. Recognizing that SLE can affect not only the subcutaneous fat layer but also the retromammary area of the breast, and understanding how it appears on breast imaging, can aid in considering this rare disease as a potential diagnosis.

Author Contributions

Conceptualization, K.W.H., K.H.J.; data curation, L.K.M., P.J.Y., N.E.J.; formal analysis, L.K.M.; methodology, L.K.M., K.W.H.; supervision, K.H.J., N.E.J.; writing—original draft, L.K.M.; and writing—review & editing, L.K.M., N.E.J., K.H.J.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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유선후층의 광범위한 석회화로 나타난 루푸스 유방염: 증례 보고

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루푸스 유방염은 전신 홍반성 루푸스의 피하지방층 침범을 의미하는 루푸스 지방층염의 일환이다. 루푸스 유방염은 드물게 발생하는 것으로 알려져 있으며 주로 유방의 만져지는 종괴로 나타난다. 전신 홍반성 루푸스를 진단받은 29세 여성이 양측 유방의 만져지는 덩이로 내원하였다. 영상 검사가 시행되었고 유방촬영술에서 광범위하고 기괴한 석회화가 주로 유선후층에서 확인되었다. 초음파에서 후방음영을 동반한 크고 불규칙한 석회화가 보였고 유방 MRI 영상에서 또한 지속적 테두리 조영증강을 보이는 광범위하고 불규칙한 병변들이 유선후층에 나타났다. 유방암을 감별하기 위해 초음파 유도하 생검을 시행하였고 루푸스 유방염의 병리적 특징인 지방 괴사를 확인하였다. 저자들은 전신 홍반성 루푸스의 매우 드문 임상양상인 루푸스 유방염의 증례를 보고하고 이 질환의 진단을 뒷받침하는 임상적, 병리적, 그리고 영상적 소견들과 감별 진단에 대해 논의하고자 한다.

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