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CASE REPORT

Lupus mastitis as an early presentation of systemic systemic lupus erythematosus

Mandana Khodashahi[1], Asal Sadat Azaminasab[1], Rozita Khodashahi[2], Soroush Sarjamee[3]

[1]*Rheumatic Disease Research Center (RDRC), Mashhad University of Medical Sciences, Mashhad, Iran*
[2]*Infectious Diseases Department, Shahid Beheshti University of Medical Sciences, Tehran, Iran*
[3]*Emergency Medicine Department, Mashhad University of Medical Sciences, Mashhad, Iran*

ABSTRACT

Lupus mastitis, as an uncommon condition in systemic lupus erythematosus (SLE), is a subset of lupus panniculitis. Here, we report a 51-year-old woman who presented with fever, weight loss, low appetite, arthralgia, and cervical lymphadenopathy. She had experienced facial palsy six months earlier to presentation. On physical examination, besides submandibular and axillary lymphadenopathy, several lumps were palpated on the breasts bilaterally. There were no abnormalities on the skin or nipples. Mammography showed diffuse calcifications in both breasts. Further work-up revealed positive anti-double stranded DNA (anti-ds DNA), antinuclear antibody (ANA), anti-Sm antibody as well as leukopenia and lymphopenia, but normal platelet count. A core biopsy of the breast showed active inflammation, leukocyte infiltration, and fat necrosis without evidence of malignancy or fibrocystic changes. Prednisone and azathioprine were initiated for her. One month later, her symptoms had improved substantially and breast ultrasound exam showed decreased size of the lumps. Although lupus mastitis is an uncommon condition, especially when it declares the onset of SLE, it should be considered in the differential diagnosis of breast lesions in lupus patients.

Key words: systemic lupus erythematosus, lupus mastitis, lymphadenopathy, biopsy

INTRODUCTION

Breast involvement in systemic lupus erythematosus (SLE), termed as lupus mastitis, is relatively uncommon. Lupus mastitis, a form of lupus panniculitis, usually involves deep subcutaneous adipose tissues of the breast. This condition can be detected by mammography. As deep tissue is affected in lupus mastitis, the condition can mimic malignancy on both clinical examination and imaging.[1-3]

Another issue that contributes to the complexity of this condition is the fact that lupus mastitis is a rare condition (with less than 50 cases reported in the literature to date) and usually is diagnosed in patients whose SLE diagnosis has been established.[3] Hence, when lupus mastitis presents at early stages of SLE without other classic and more well-known criteria for SLE, its diagnosis could be challenging. In addition to imaging, histologic examination plays a crucial role in the diagnosis of lupus mastitis. Although features of lupus mastitis are often confused with malignancy, lymphoma, and tuberculosis, its histologic appearances are distinct.[4]

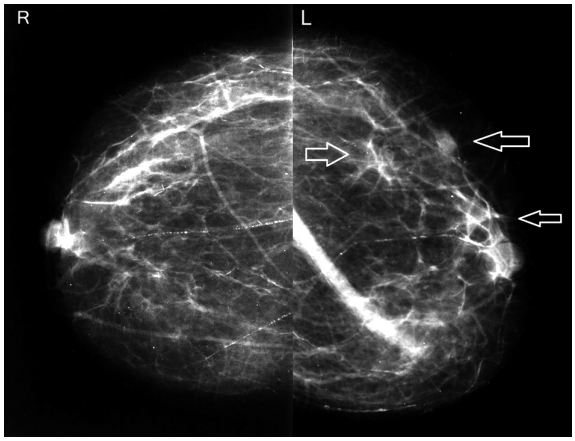

Figure 1, Right (R) and left (L) mammograms show focal asymmetry at the upper outer quadrant position of the left breast (L) and heterogeneous vascular calcifications in both breasts (arrows).

Here, we present an unusual case of lupus mastitis in a middle-aged woman which occurred earlier than other most well-known manifestations of SLE.

Correspondence:
Mandana Khodashahi, MD
Rheumatic Disease Research Center (RDRC), Ghaem Hospital,
Ahmad-Abad Ave., Dr Shariati Sq.,
Mashhad, Razavi Khorasan, Iran
Tel: +98 513 8012753
E-mail: khodashahimn@mums.ac.ir

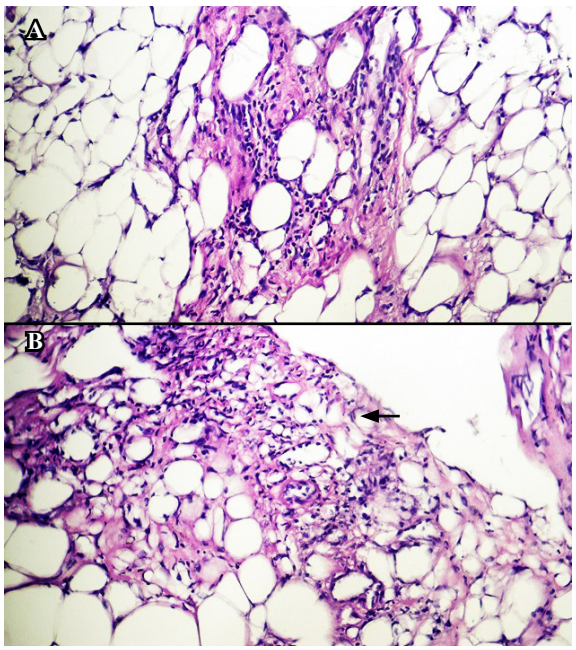

Figure 2, Histopathologic examination of the breast sample biopsy shows leukocyte infiltration and fat necrosis (A) as well as active inflammation and necrosis of adipose tissue (B) (arrow) without evidence of malignancy or fibrocystic changes (H&E × 400).

CASE REPORT

A 51-year-old woman, who had history of facial palsy several months earlier, was admitted to our hospital due to fever, weight loss, low appetite, and arthralgia for the past six months. Physical examination showed lymphadenopathy in the cervical, submandibular, and axillary regions. The largest lymphadenopathies in the axillary regions measured 48 × 14 mm in size in the left side and 46 × 18 mm in the right side. Plus, several lumps were palpated on breasts. However, skin and nipple examination were unremarkable and no sign of skin change, dimpling, or ulcer were found. The presumptive diagnoses of an infectious process (such as tuberculosis), lymphoma, malignancy (in particular breast malignancy considering the palpated lumps), and connective tissue diseases were made.

Laboratory examination showed leukopenia (3.4 × 103 per cc) and lymphopenia (17%) with normal hemoglobin and platelet levels, elevated ESR (60 mm/h), elevated hs-CRP (12 mg/L) and low levels of complement proteins (C3= 28 mg/dL (normal range= 90-180 mg/dL) and C4= 1 mg/dL with normal range of 10-40 mg/dL). Anti-double stranded DNA (anti-ds DNA) was 429 U/mL (normal < 100 U/mL) and antinuclear antibody (ANA) was 6.5 IU (normal < 1 IU). Anti-Smith (SM) antibody was also positive. Other laboratory tests including urinalysis, serum creatinine, direct Coombs test, anti-phospholipid antibodies (anti-cardiolipin antibodies, lupus anti-coagulant antibody, beta-2 glycoprotein-1 antibody), CMV ELISA antibodies, TB PCR, and CMV PCR on cervical lymph node biopsy were within normal range.

On ultrasound exam, the largest lymph node was in the right submandibular gland, which measured 9×5×6 mm; all nodes were reactive lymph nodes with normal shape and echogenicity. Breast ultrasound described the presence of some heterogeneous masses, the largest measured 22 × 8 mm. The masses were hypoechoic with defined margins suggestive of fibroadenoma or hamartoma. Abdominal and pelvic ultrasound examinations as well as trans-thoracic echocardiography were unremarkable. Biopsy of the enlarged lymph nodes of the cervical and axillary regions showed reactive follicular hyperplasia without tumoral tissue.

Considering the clinical and laboratory findings including facial palsy, arthralgia, weight loss, leukopenia, lymphopenia, low levels of complement proteins, positive anti-ds DNA and ANA, the likely diagnosis of SLE was made. However, the patient did not have other more classic clinical manifestations of SLE such as photosensitivity, malar rash, or oral ulcers. Due to the history of idiopathic peripheral facial nerve (the 7th cranial nerve) palsy, neurological examination was done which was normal. MRI of the brain showed the pattern of small vessel disease which was related to senile atherosclerosis. Thus, it seemed that her facial palsy is also a manifestation of underlying SLE.

Because breast masses were peculiar in the clinical setting, more accurate examination of breast masses was performed by mammography. It showed focal asymmetry at the upper outer quadrant position of the left breast with diffuse calcifications in both breasts (Fig. 1). A core biopsy of the breast mass was performed which characterized fat lobules with focal fat inflammation and necrosis without evidence of malignancy or fibrocystic changes (Fig. 2). These features were compatible with the diagnosis of lupus mastitis, a feature of lupus panniculitis.

Treatment was done based on immunosuppressive therapy by administering prednisone (40 mg/day) and azathioprine (2 mg/kg). On follow-up evaluation performed one month later, her symptoms had improved and WBC, ESR, hs-CRP, anti-ds DNA, and complement proteins levels were found to be normal. On ultrasound evaluation, there was a moderate decrease in the size of the breast lumps.

DISCUSSION

Lupus panniculitis is a rare clinical variant of SLE which occurs when the deep dermis and subcutaneous fat are predominantly affected.[5] LP is an infrequent manifestation of SLE, occurring in only 2% to 3% of SLE patients.[4] If breast tissue is involved, lupus panniculitis is termed lupus mastitis. Fewer than 50 cases of lupus mastitis have been reported so far and most of them are female.[4]

Although lupus mastitis affects patients which are usually known to have SLE or discoid lupus erythematosus (DLE), it can also herald the onset of SLE or DLE.[6] Our patient presented with adenopathy, fever, arthralgia, breasts masses and no prior diagnosis

of SLE. Sonography showed the echogenic nature of the masses in the breasts, an appearance that can suggest malignancy. Features of axillary lymph nodes suggested a systemic disease without relation to the breast masses. Due to the wide lymphadenopathies detected in the neck and axillary regions and because of possibility of neoplastic processes and other inflammatory disorders, more investigations were carried out for making an accurate diagnosis.

On biopsy examination of the lymph node specimens, lymph nodes of the neck masses showed reactive follicular hyperplasia without malignancy. Result of breast biopsies also indicated presence of fat inflammation and necrosis without tumor involvement. On the other hand, our patient had a history of facial palsy 6 months ago. Neurologic manifestations have been reported during the course of the disease and rarely the first manifestation of SLE.[7] Positive ANA titer and anti-ds DNA assay in this patient also helped to connect the reason of facial palsy to SLE.

Overall clinical tests, biopsies, and history of facial palsy were strongly suggestive of lupus mastitis although the history of SLE did not exist.

CONCLUSION

Although lupus mastitis is an uncommon condition, especially when it declares the onset of SLE, it should be considered in the differential diagnosis of breast lesions in lupus patients. The lymphocytic inflammation and fat involvement of adipose tissue in the breast could be the first clue that may aid in differentiating lupus mastitis from lymphoma or malignancy. In this case, facial palsy, the rare manifestation of SLE, also was evident in a middle-aged woman which helped to diagnose SLE.

CONFILICT OF INTERESTS

None.

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