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Case Report

Lupus Mastitis with Cardiopulmonary Involvement in Overlap Syndrome

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Abstract:

Systemic lupus erythematosus (SLE) is a complex autoimmune disorder with multisystem involvement, including rare variants like lupus panniculitis (LP). This variant is characterized by chronic inflammation of subcutaneous fat. Lupus mastitis (LM), an uncommon manifestation of LP, involves the breast's deep subcutaneous fatty tissue and presents usually as a diagnostic challenge. We describe a healthy young lady in her mid-twenties who presented with bilateral breast pain and swelling. Further evaluation revealed lupus mastitis as the initial manifestation of an overlap syndrome involving systemic lupus erythematosus and polymyositis, with associated cardiopulmonary involvement. She experienced several symptoms including proximal muscle weakness, and respiratory symptoms. Following immunosuppressive therapy with corticosteroids, mycophenolate mofetil, hydroxychloroquine, and rituximab, she showed significant clinical improvement across all affected systems. This case highlights the importance of thorough evaluation in patients presenting with breast swelling, emphasizing the need to avoid unnecessary surgical interventions, and underscores the effective management strategies for complex cases of LM associated with overlap syndrome.

Key words: lupus mastitis; overlap syndrome; polymyositis; pulmonary hypertension; breast swelling

Introduction

Systemic lupus erythematosus (SLE) is a disease that affects various body systems. It predominantly impacts women in their reproductive age. It can manifest in different system including skin, kidney and other organs.

Lupus panniculitis (LP), also known as lupus profundus, is a chronic inflammatory condition of the deep dermis and subcutaneous fat associated with systemic or cutaneous lupus erythematosus. It typically presents as firm, tender subcutaneous nodules, often with overlying skin changes such as atrophy or hyperpigmentation. This process can involve several tissues. When it involves the breast, it is termed as lupus mastitis (LM), a rare and localized variant of LP that may clinically mimic infectious or malignant processes, making diagnosis challenging.[1]

Lupus mastitis (LM) is a form of lupus panniculitis that affects around 2-3% of individuals with lupus. It is more prevalent among women of African backgrounds. While its exact cause remains, unknown LM is believed to be a condition resembling other manifestations of SLE. It is usually characterised with inflammation in the adipose tissue of the breast accompanied by significant plasma cell and lymphocytes infiltration, and necrosis of adipose lobules [1-3]. The aim of the current article is to report a rare case of lupus mastitis as the initial manifestation of an overlap syndrome involving systemic lupus erythematosus and polymyositis, with cardiopulmonary involvement, and to highlight the importance of early diagnosis and management.

Case Presentation:

A young adult female in her mid-twenties without any existing health conditions, presented to rheumatology service with one month history of pain, swelling and hardness in both breasts. She mentioned experiencing fever, nausea, vomiting and abdominal pain before coming to the hospital. Additionally, she complained of weakness in her legs with general joints pain and occasional episodes of unconsciousness without convulsions or unusual movements. However, she did not display any skin rashes, mouth ulcers or Raynaud's phenomenon. She also described feeling fatigue and overall body weakness. The patient had trouble getting up from a seated position and climbing stairs. During the exam the patient was alert and oriented to time, place and person. Her vital signs were normal with no signs of enlarged lymph nodes. Breast examination revealed hardening, induration and tenderness in both breasts, but no changes in the overlying skin or any discharge. Lung examination showed clear breath sounds in both lung fields. There was no oedema in her lower limbs. Muscle examination indicated mild tenderness and weakness in both upper and lower limbs (4/5), more in the thigh muscles without any signs of muscle wasting or atrophy. There was no evidence of any active

synovitis or skin rash in her clinical examination. The lab tests showed that the haemoglobin level was 9.8 g/dL which is, below the range of (12 -16 g/ l), White blood cells were at 10.3 x 10^9/L (4.0-11.0 x 10^9g/L), and platelet count was within normal limits at 449 x 10^9/L (150 - 450 x 10^9 g/L). Direct Antiglobulin Test came as positive. Liver function tests indicated an increased ALT level of 60 IU/L (40-50 IU/L) and a decreased albumin level of 20 g/L (35 -50 g/L). Renal function appeared normal. Her Inflammatory markers were notably high with a CRP value of 140 mg/L (Normal <5 mg/L) and ESR exceeding130 mm/h. The levels of complement were normal, with C3 at 1531 mg/L (850 - 1600 mg/L) and C4 at 229 mg/L (120 -360 mg/L). Muscle enzymes showed an increase with CK at 726 IU/L (34 -145 IU/L) and myoglobin >1000 µg/L (25 - 58 µg/L). Her viral serology work up came as negative for Hepatitis B surface antigen, core antibodies and for Hepatitis C antibodies, as well as nonreactive HIV tests (1 & 2). The patients IgG subclass 4 level was measured at 0.407 g/L (range; 0.040- 0.870 g/L) and her angiotensin converting enzyme (ACE) level was recorded as 33 U/L (20-70 U/L). The quantiferon TB Gold test was negative as well.

In the serology and autoimmune panel Anti-Nuclear Antibodies (ANA) were positive, showing reactivity to Anti RNP/Sm and Anti SSA (Ro) antibodies, while other antibodies, like Anti dsDNA, Anti Jo 1, Anti Scl 70 tested as negative. Myositis specific antibodies screen panel came as negative. The initial investigation for antiphospholipid antibodies showed positive anticardiolipin IgG. However, tests for lupus anticoagulant and beta 2 glycoprotein came back as negative. Subsequent tests after 12 weeks, repeated antiphospholipid antibodies showed that; the positive anti cardiolipin IgG had reverted to negative excluding the presence of persistent antiphospholipid antibodies. Several radiological imaging was conducted to assess the complex nature of the patient presentation, included breast ultrasounds, which identified small anechoic cysts, suggestive of simple cysts, with no masses or inflammatory changes (BIRADS 2). Thigh MRI was conducted to assess the extent of muscle involvement and it showed diffuse symmetrical muscular oedema with subcutaneous fatty oedema and postcontrast enhancement in the gracilis muscles. Additionally, she had a mammogram done and it showed no suspicious lesions.

Patient underwent PET/CT scan was performed to rule out any underlying malignancy and the report revealed diffuse symmetrical FDG activity within all skeletal muscles, suggestive of myositis, and mild heterogeneous thickening of the gallbladder, with no evidence of malignancy. **Figure 1**

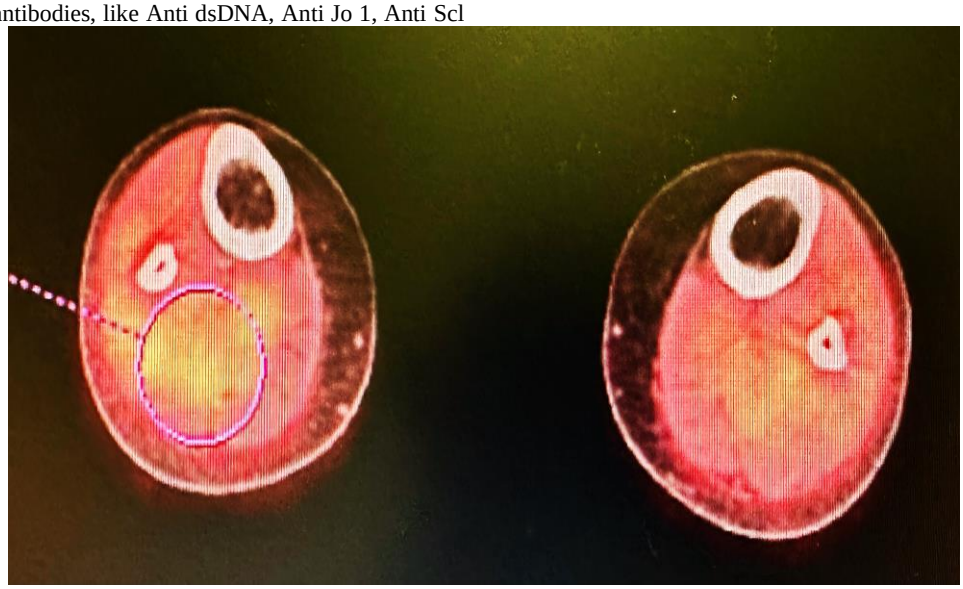

Figure 1: This PET scan shows axial cross-sections of the skeletal muscles with diffuse symmetrical FDG uptake, indicating widespread muscle inflammation. The highlighted region on the left denotes an area of increased metabolic activity, consistent with inflammatory myopathy.

The initial echocardiogram revealed significant findings, including a dilated right ventricle with systolic dysfunction and moderate tricuspid regurgitation. The estimated pulmonary pressure was high approximately 48 mmHg. There was also mild to moderate pericardial effusion, but the left ventricular size and function were preserved, with a normal left ventricular ejection fraction. These findings suggested increased pulmonary artery pressures with pericardial effusion secondary to the underlying inflammatory condition. She had a pulmonary CT angiogram to rule out pulmonary embolism in view of raised pulmonary artery pressure in the echocardiogram and it showed a dilated right atrium and ventricle, accompanied by moderate pericardial effusion. Cardiac CT showed dilation of the right atrium and ventricles, while the remaining cardiac structures appeared unremarkable.

Patient also underwent a core biopsy of the breast tissue which revealed benign fibroadipose tissue with fat necrosis and neutrophilic infiltration, consistent with lupus mastitis and with no evidence of malignancy.

The diagnosis of overlap syndrome lupus/polymyositis was considered in the presence of polymyositis, lupus mastitis and cardiopulmonary involvement. Consequently, the patient was initially treated with intravenous methylprednisolone, Mycophenolate mofetil, hydroxychloroquine, and Rituximab. The patient had substantial clinical improvement with a reduction of the breast induration and soreness over time. Furthermore, she regained her muscular strength. The patient's repeated blood work up showed improvement in her muscle enzymes (Creatine Kinase: 28 IU/L, Myoglobin: 12 µg/L). In addition, there was a decrease in inflammatory markers (CRP: 14 mg/L, ESR: 20 mm/hr). **Table 1**

Period	ESR (mm/hr)	CRP (mg/L)	CK (IU/L)	LDH (IU/L)	Myoglobin (µg/L)
At presentation	>130	140	726	398	617
Two weeks	74	-	217	464	373
I month	22	<4	84	325	94

3 months	40	8	29	-	17
6 months	25	<0.4	25	182	15
1 year	-	14	28	197	12

Table 1: Longitudinal laboratory monitoring of the patient over time

The Liver function tests were stable (ALT: 21 IU/L, AST: 22 IU/L, Albumin: 35 g/L). The levels of complement were within the normal range, with C3 at 1477 mg/dL and C4 at 264 mg/dL. Additionally, a follow-up echocardiography verified the absence of notable pulmonary hypertension, which reflect a favourable response to therapy. The patient is currently maintained on Mycophenolate Mofetil and Hydroxychloroquine. She was advised to continue regular follow-ups with cardiology and rheumatology teams to assess her disease progression.

Discussion

Lupus mastitis, also known as lupus erythematosus profundus is a form of lupus panniculitis where inflammation occurs in the fatty tissue beneath the skin of the breast. This condition is part of a group of diseases that includes systemic lupus erythematosus and discoid lupus erythematosus (DLE). Initially it was described by Kaposi in 1883. Lupus mastitis typically affects a small percentage of lupus patients, mostly adult women aged between 20 to 60 years old, though cases have been reported in younger patients and occasionally in men. [4-6]

The accurate diagnosis of lupus mastitis involves a combination of the following elements: detailed medical history, diagnostic laboratory work-up, radiological imaging, and analysis of tissue biopsy. It is essential to understand the clinical and radiological characteristics associated with LM to prevent misdiagnosis and unnecessary surgical procedures. [7]

In general, lupus mastitis presents as nodules under the skin that may or may not cause pain. The affected area may show skin changes like thinning, redness or darkened pigmentation. In our case study, the patient experienced pain and swelling in both breasts without any noticeable changes on the skin surface. The results of a mammogram may show densities and calcifications, or bright masses on ultrasound images, which require a careful assessment to distinguish it from other conditions like inflammatory breast cancer and diabetic mastopathy. [8-9]

Lupus mastitis is identified by the infiltration of lymphocytes in the lobules along with hyaline fat necrosis and fibrosis. The presence of panniculitis is also a distinctive feature. Calcifications are commonly observed in more advanced stages. These specific tissue characteristics, combined with the patient's background play a crucial role in distinguishing lupus mastitis, from malignancies and other inflammatory breast disorders. [10-11]

It worth mentioning, that fat necrosis in the breast can arise from various other causes, including surgical procedures, trauma, radiation therapy, autoimmune and inflammatory conditions. Compromised blood supply can lead to tissue necrosis like in the presence of infections and vasculitis. This mechanism can manifest in breast tissue and cause fat necrosis. Furthermore, fat necrosis can occur from simple mechanical factors such as tight garments, or develop spontaneously without specific cause, a condition known as idiopathic fat necrosis. Therefore, distinguishing different mechanisms that had led to fat necrosis is crucial for accurate diagnosis and appropriate early management. [8,9].

The differential diagnosis lupus mastitis covers a wide range of conditions with similar symptoms, including infections, granulomatous and infiltrative breast conditions, inflammatory breast cancer, diabetic mastopathy, and vasculitis. Inflammatory breast cancer is a special consideration due to similar skin changes and the presence of a palpable lump. Lupus mastitis is identified by the absence of malignant cells and the presence of specific histological features like hyaline fat necrosis and fibrosis. Diabetic mastopathy on other hand shows fibrosis but lacks the autoimmune features seen in lupus. While rare, primary medullary carcinoma can also appear as breast masses, and it can be confirmed as malignant through histological examination. [7,10-13].

The main approach in treating lupus mastitis includes the use of medications such as hydroxychloroquine in combination with corticosteroids to decrease inflammation and effectively control the advancement of the disease. In cases where the condition is sever, additional immunosuppressive drugs, like methotrexate, cyclophosphamide or rituximab may be required. Surgical procedures are generally not preferred as they have the potential to worsen mastitis. If biopsies are needed non-invasive techniques should be utilized. [11,14-19] **Table 2**

Case	Reference	Patient Age and Gender	Treatment Received
1	Carducci et al., 2005[6]	62-year-old female	Antimalarial therapy
2	Chen et al., 2005[11]	29-year-old female	Immunosuppressive therapy
3	Fernandez-Flores et al., 2006[12]	42-year-old male	(No specific treatment reported)
4	Bachmeyer C., 2006[19]	30-year-old female	Steroids, antimalarial drugs
5	Nigar et al., 2007[13]	40-year-old female	(No specific treatment reported)
6	Bayar et al., 2007[17]	23-year-old female	Suggested medical treatment with steroids or antimalarial drugs
7	Summers et al., 2009[7]	43-year-old female, in addition to 20 other cases.	Suggested treatment options included hydroxychloroquine, systemic steroids at a dosage of 1 mg/kg/day, and cyclophosphamide.
8	Guerre et al., 2009[5]	46-year-old female	Antimalarials, corticosteroids, immunosuppressants
9	Wang et al., 2010[18]	28-year-old female	Corticosteroids
10	Oktay et al., 2021[2]	37-year-old female	Steroids, antimalarial drugs
11	He IJ et al., 2023[14]	48-year-old female	Anticoagulation, rituximab, mycophenolate mofetil, quinacrine
12	Yong Guo et al., 2023[16]	27-year-old female	Systemic corticosteroids, hydroxychloroquine
13	Mazeda et al., 2023[4]	34-year-old female	Hydroxychloroquine, prednisolone, rituximab
14	Wang et al., 2024[1]	26-year-old female	Antimalarial (hydroxychloroquine), systemic steroid therapy
15	**[Current case, 2024]**	26-year-old female	Steroids, mycophenolate mofetil, hydroxychloroquine, rituximab

Table 2: Treatment Approaches for Lupus Mastitis in Various Case Studies

The current presented case demonstrates a significant complexity due to the presence of overlap syndromes involving both lupus and polymyositis. The patient experienced cardiac and pulmonary complications, including pericardial effusion and high pulmonary

pressure. Fortunately, early initiation of therapy had led to remarkable improvement across all symptoms as confirmed by subsequent evaluation. The successful treatment and recovery underscore the importance of early intervention in such instances.

Conclusion:

This case highlights the significance of conducting a thorough assessment when evaluating patients presenting with breast mass and swelling before considering any surgical interventions. It stresses the importance of diagnosing conditions such as lupus mastitis which can resemble cancer but do not necessarily require surgical intervention like mastectomy. By identifying various differential diagnosis that can cause such conditions, healthcare providers can ensure patients receive appropriate treatments thereby and improving overall outcomes. Researchers should delve into identifying genetic and epigenetic factors that play important role in the development of lupus mastitis (LM) among SLE patients.

Declaration of Conflicting Interests:

The authors declare that there are no conflicts of interest related to the research, authorship, or publication of this article.

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