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FROM BREAST MASS TO PARALYSIS: LYMPHOCYTIC MASTITIS AS THE SENTINEL MANIFESTATION OF SJÖGREN'S SYNDROME

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ABSTRACT

Sjögren's syndrome is a systemic autoimmune disease classically characterized by sicca symptoms; however, extra glandular manifestations may precede glandular involvement and, in rare cases, present as life threatening emergencies. We report a middle-aged patient who presented with acute flaccid paralysis due to severe hypokalaemia secondary to distal renal tubular acidosis, requiring intensive care admission. Notably, the patient had undergone breast excision two years earlier for a painful breast mass, with histopathology highly suggestive of lymphocytic mastitis, which was misdiagnosed and treated as a breast abscess. Subsequent immunological evaluation confirmed primary Sjögren's syndrome. Prompt electrolyte correction and immunosuppressive therapy resulted in complete neurological recovery.

KEYWORDS

Sjögren's syndrome, lymphocytic mastitis, hypokalaemic paralysis, distal renal tubular acidosis, autoimmune disease

LEARNING POINTS

- Sjögren's syndrome may initially present with rare extra glandular manifestations, preceding classical sicca symptoms and leading to significant diagnostic delay.
- Lymphocytic mastitis can represent an early autoimmune manifestation of Sjögren's syndrome and may closely mimic infectious or malignant breast disease.
- Distal renal tubular acidosis is the most frequent renal manifestation of Sjögren's syndrome and may result in life-threatening hypokalemia and acute flaccid paralysis.

INTRODUCTION

Sjögren's syndrome (SS) is a chronic systemic autoimmune disease characterized by lymphocytic infiltration and progressive dysfunction of exocrine glands, particularly the salivary and lacrimal glands, resulting in xerostomia and xerophthalmia[1,2]. Although sicca symptoms are considered the hallmark of SS, up to one-third of patients develop extra glandular manifestations involving the kidneys, nervous

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system, lungs, skin, and musculoskeletal system. These systemic features may precede or overshadow glandular symptoms, often leading to diagnostic delay[3,4].

Renal involvement is reported in approximately 5–14% of patients with SS and is most commonly due to immune-mediated tubulointerstitial nephritis[5]. Distal renal tubular acidosis (dRTA) represents the most frequent renal manifestation and results from impaired hydrogen ion secretion in the distal tubules due to lymphocytic infiltration and tubular damage[6]. Clinically, dRTA may cause metabolic acidosis, nephrolithiasis, and severe hypokalemia. Profound hypokalemia can manifest as muscle weakness, paralysis, cardiac arrhythmias, and, in rare cases, life-threatening hypokalaemic periodic paralysis[7].

Lymphocytic mastitis, also known as diabetic or autoimmune mastitis, is a rare benign inflammatory breast condition characterized histologically by dense periductal and perivascular lymphocytic infiltration with stromal fibrosis. Although most commonly described in patients with long-standing diabetes mellitus, lymphocytic mastitis has also been reported in association with autoimmune diseases, including SS, systemic lupus erythematosus, and Hashimoto's thyroiditis[8]. Owing to its clinical and radiological resemblance to infectious abscesses or breast malignancy, it is frequently misdiagnosed, resulting in unnecessary surgical intervention.

This case highlights lymphocytic mastitis as a rare and under-recognized early extra glandular manifestation of SS, preceding the diagnosis by several years and culminating in a life-threatening presentation of hypokalaemic paralysis secondary to dRTA.

CASE DESCRIPTION

A 47-year-old Saudi woman with no previously diagnosed autoimmune disease apart from Hashimoto's hypothyroidism, treated with oral levothyroxine 125 µg once daily, presented with an acute onset of generalized weakness and numbness that rapidly progressed to flaccid paralysis. On examination, her vital signs were stable. Systemic examination was unremarkable except for a malar rash, reduced salivary pooling with tongue fissuring *(Fig. 1)*, and purpuric skin rash over the lower and upper limbs *(Fig. 2)*. Neurological examination revealed a Glasgow coma scale score of 15/15 with intact cranial nerves, reduced muscle tone, and symmetric proximal and distal muscle weakness with a power of 3/5 in both upper and lower extremities.

An electrocardiogram performed in the emergency department demonstrated a prolonged PR interval with prominent U waves. Initial laboratory investigations revealed severe hypokalemia (serum potassium 2.0 mmol/l), normal anion gap metabolic acidosis (anion gap 12 mmol/l), arterial pH of 7.24, and bicarbonate level of 17.3 mmol/l. Urinary studies showed an inappropriately high urine pH of 7, 24-hour urinary protein excretion of 1 g/day, and an elevated trans tubular potassium gradient of 11, supporting the diagnosis of distal renal tubular acidosis (type 1) *(Tables 1 and 2)*.

The patient was admitted to the intensive care unit with a diagnosis of hypokalaemic periodic paralysis for close monitoring and correction of severe hypokalemia. She received intravenous potassium chloride replacement, resulting in rapid clinical improvement, and was transferred to the general ward after one day in a stable condition with a serum potassium level of 3.2 mmol/l.

Further history revealed a two-year history of sicca



Figure 1. *Oral mucosal involvement at presentation. Clinical photograph demonstrating a dry, erythematous tongue with fissuring, consistent with xerostomia and oral mucosal involvement as an extra glandular manifestation of Sjögren's syndrome.*



Figure 2. *Cutaneous vasculitic rash of the upper limb. Photograph showing diffuse erythematous maculopapular rash with mottled appearance over the forearm and hand, consistent with vasculitic skin involvement associated with systemic autoimmune disease.*

Parameter	Result	Reference range
Hb (g/dl)	10.8	12-16
WBC count (×10³/µl)	4.71	4-11
Platelets (×10³/µl)	406	140-450
CRP (mg/l)	9.2	≤5
ESR (mm/h)	119	0-20
Serum potassium (mmol/l)	2	3.5-5.2
Serum sodium (mmol/l)	134.93	36-145
Serum chloride (mmol/l)	105	96-106
Serum bicarbonate (mmol/l)	17.3	22-26
Venous pH	7.24	7.32-7.42
Serum creatinine (µmol/l)	80	49-90
Blood urea Nitrogen (mmol/l)	4.6	2.5-6.4
Anion gap (calculated) (mmol/l)	12	4-12

Table 1. Baseline laboratory investigations on admission. Abbreviations: Hb, haemoglobin; WBC, white blood cell; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate.

symptoms, including dry eyes and dry mouth requiring frequent water intake to aid swallowing, in addition to hair thinning, photosensitive facial rash, and unintentional weight loss of approximately 20 kg over one year.

An extensive diagnostic evaluation was performed to rule out secondary causes of hypokalemia including malignancy. Serological testing, labial salivary gland biopsy, skin biopsy, bone marrow aspiration, pan-computed tomography, colonoscopy, and gastroscopy were performed. A diagnosis of primary Sjögren's disease was established based on positive antinuclear antibodies, anti-Ro/SSA, and anti-La/SSB antibodies, along with supportive histopathological findings on labial salivary gland biopsy *(Table 3)*. Skin biopsy revealed leukocytoclastic vasculitis, consistent with immune-complex–mediated small-vessel inflammation. The nephrology team deferred renal biopsy as 24- hour urinary protein excretion normalized following corticosteroid therapy. All other investigations were unremarkable.

The patient demonstrated marked clinical improvement following potassium supplementation, alkali therapy, and systemic corticosteroids, with complete resolution of neurological deficits *(Table 4)*.

Notably, two years prior to the current admission, the patient had been hospitalized under the surgical service for a painful breast mass of more than five years' duration. She underwent breast excision, and histopathological examination revealed features highly suggestive of lymphocytic mastitis, characterized by prominent periductal fibrosis and dense lymphocytic infiltration. Due to delayed immunological evaluation and overlapping clinical and radiological features, the condition was managed as a breast abscess, and no autoimmune workup was pursued. In retrospect, this presentation likely represented an early extra glandular manifestation of SS and a diagnostic mimicker of infectious breast disease.

The patient continues regular follow-up at the rheumatology clinic and remains on hydroxychloroquine 400 mg per orally once daily, mycophenolate mofetil 500 mg orally once daily, and cyclical rituximab every six months with sustained clinical improvement. Corticosteroids were gradually tapered and ultimately discontinued.

DISCUSSION

This case illustrates an unusual and instructive sequence of extra glandular manifestations of SS, beginning with lymphocytic mastitis and later progressing to severe hypokalaemic paralysis caused by distal renal tubular acidosis. Such a temporal pattern emphasizes the heterogeneous nature of SS and the potential for extra glandular involvement to precede classical sicca symptoms by several years[3].

Lymphocytic mastitis is an uncommon inflammatory breast condition characterized by periductal and lobular lymphocytic infiltration with associated fibrosis. Its association with autoimmune disease is increasingly recognized but remains poorly appreciated in clinical practice. Several case reports have described lymphocytic

Parameter	Result	Reference
Urine pH	7	4.5-8 (normal renal acidification in metabolic acidosis: <5.5)
Urine potassium (mmol/l)	18.6	25-125 (spot urine)
Urine sodium (mmol/l)	70.70	20-220 (spot urine)
Urine chloride (mmol/l)	50	20-250 (spot urine)
Urine anion gap (mmol/l)	39.3	<0
Trans tubular potassium gradient	11	<3 in hypokalemia

Table 2. Urine studies and acid–base evaluation.

Test	Result	Reference range
ANA	>1:1280 speckled	<1:80
Anti dsDNA	Negative	<1:10
Anti-Ro / SSA	Positive	Negative
Anti-La / SSB	Positive	Negative
Rheumatoid Factor	>200	<30
Complement C3	1.47	0.83-1.93
Complement C4	0.386	0.15-0.57

Table 3. Immunological and autoimmune profile.
Abbreviations: ANA, antinuclear antibodies; dsDNA, double stranded deoxyribonucleic acid.

Parameter	Upon admission	Post-treatment
Serum potassium (mmol/l)	2	3.2
Serum bicarbonate (mmol/l)	17.3	22.2
Venous pH	7.24	7.35
Muscle strength	3 out of 5	5 out of 5
Serum creatinine (µmol/l)	80	66

Table 4. Follow-up laboratory results after treatment.

mastitis in patients subsequently diagnosed with SS or other systemic autoimmune disorders, suggesting that it may represent an early localized immune manifestation rather than an isolated breast pathology[9]. In the present case, the absence of infectious risk factors and the histopathological findings should have prompted an autoimmune evaluation. Failure to recognize its immune-mediated nature resulted in delayed diagnosis of SS.

Renal involvement in SS is most commonly related to tubulointerstitial nephritis, which disrupts distal tubular acidification and leads to dRTA[6]. Hypokalemia results from renal potassium wasting secondary to impaired hydrogen ion secretion. Severe hypokalemia may manifest dramatically as acute flaccid paralysis, as observed in this patient. Similar cases of hypokalaemic paralysis as the initial presentation of SS have been reported, highlighting the importance of considering autoimmune aetiologies in patients presenting with unexplained electrolyte disturbances[7]. Neurological deficits are typically reversible with timely potassium replacement, alkali therapy, and immunosuppressive treatment.

Cutaneous vasculitis is a well-recognized extra glandular manifestation of SS and most commonly presents as palpable purpura involving the lower extremities. Histopathologically, leukocytoclastic vasculitis reflects immune-complex–mediated small-vessel inflammation and is associated with higher systemic disease activity, hypocomplementemia, and multiorgan involvement[4,10]. In this patient, biopsy-proven leukocytoclastic vasculitis further supports the systemic nature of the disease and underscores the importance of considering SS in patients presenting with multisystem inflammatory features, even in the absence of prominent sicca symptoms.

Overall, this case emphasizes the need for a high index of suspicion for autoimmune disease in patients presenting with atypical inflammatory conditions affecting multiple organ systems over time. Early recognition of rare extra glandular manifestations, such as lymphocytic mastitis, may facilitate timely diagnosis, prevent serious complications, and improve long-term outcomes in patients with SS.

CONCLUSION

Lymphocytic mastitis may represent a sentinel extra glandular manifestation of SS. Failure to recognize autoimmune mimickers can result in delayed diagnosis and severe complications. Early consideration of systemic autoimmune disease in unexplained inflammatory breast lesions and hypokalaemic paralysis is crucial to ensure timely diagnosis and favourable outcomes.

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