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Nephrocalcinosis in Sjögren Syndrome - Case Report

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Abstract

Sjögren's syndrome is defined as an immunologic disorder characterized by progressive lymphocytic destruction of exocrine glands, most frequently leading to symptomatic eye and mouth dryness. It is a chronic systemic autoimmune disease that can affect other organs and tissues, resulting in both glandular (sicca symptoms) and extraglandular manifestations. It predominantly affects women (≈9:1) with onset in the fourth to sixth decades of life and may occur as primary disease or in association with other rheumatologic conditions such as rheumatoid arthritis or systemic lupus erythematosus. Pathogenesis involves CD4⁺ T-cell–mediated glandular destruction, B-cell hyperactivity with production of autoantibodies (anti-Ro/SSA and anti-La/SSB), and cytokine-driven dysfunction; persistent infections (e.g., Epstein-Barr virus) and genetic predisposition contribute to disease expression. Clinical presentation includes sicca symptoms (dry eyes, dry mouth), parotid enlargement, and extraglandular manifestations such as arthritis, pulmonary involvement, renal tubular acidosis, neuropathy, and heightened lymphoma risk. Diagnosis is based on objective ocular and salivary tests (Schirmer's test, unstimulated salivary flow), labial salivary gland biopsy demonstrating focal lymphocytic sialadenitis (focus score ≥1/4 mm²), and serology for anti-SSA/Ro or anti-SSB/La antibodies, often using ACR/EULAR criteria with a point-based scoring system. Prognosis varies with extraglandular involvement; vigilant monitoring for lymphoma and organ-specific complications is essential.

Nephrocalcinosis in sjögren syndrome

Sjögren syndrome is a systemic autoimmune disorder most commonly presenting with sicca symptoms. Sicca refers to dryness most often involving the eyes and mouth due to inflammation and resultant pathology of the lacrimal and salivary glands. Up to one-half of affected individuals also develop extra-glandular involvement implying the occurrence of signs and symptoms in organs distinct from the salivary and lacrimal glands including the joints, skin, lungs, Gastrointestinal (GI) tract, nervous system, and kidneys. Sjögren syndrome frequently occurs in association with other autoimmune disorders including Rheumatoid Arthritis (RA) and Systemic Lupus Erythematosus (SLE). In the early 1900s, Swedish physician Henrik Sjögren (SHOW-gren) first described a group of women whose chronic arthritis was accompanied by dry eyes and dry mouth. Although Sjögren syndrome is classically considered to be localized disease of the exocrine glands, mainly manifested with

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oral and ocular dryness, it also has a wide range of systemic clinical manifestations that affect essentially any organ system, and a small number of cases are complicated by the development of non-Hodgkin lymphoma. The prevalence of primary Sjögren's syndrome is ~0.5–1%, while 5–20% of patients with other autoimmune diseases suffer from Sjögren's syndrome (secondary). It is the most common autoimmune disease after rheumatoid arthritis (Carsons and Patel, 2021; Qin et al., 2015). There is a higher prevalence of the disease in Caucasian populations (particularly in European cohorts) and a lower prevalence in Hispanic populations (Brito-Zerón et al., 2017). Sjögren's syndrome is likely underdiagnosed either because a physician does not recognize the features of this systemic condition or the patient does not feel that it is necessary to visit the doctor for dry eyes and mouth (Sandhya et al., 2017). Renal involvement is uncommon (less than 10 percent) and includes a spectrum of manifestations, interstitial nephritis being the most prevalent. Electrolyte disturbances can develop, most notably distal renal tubular acidosis, but diabetes insipidus and Gitelman syndrome (François and Mariette, 2016) are also possible. End-stage renal disease is rare (less than 1 percent) (Lin et al., 2020). There is a well-established association between primary Sjogren's syndrome and distal renal tubular acidosis (dRTA). Drta is a relatively infrequent manifestation of primary Sjögren's syndrome which can present with life-threatening electrolyte abnormalities while, in some patients, it could be the first manifestation of the syndrome. The reported prevalence of renal involvement in SS varies widely, ranging between 2% and 67%Anemia is present in 20 percent of SS cases, usually normochromic or normocytic. Leukopenia is found in 15 percent of patients and corresponds to lymphocytopenia. CD4-lymphocytopenia is mainly found in anti-Ro-SSA positive patients and is associated with an increased risk of non-Hodgkin's lymphoma (Parisis et al., 2020).

Case report

A 40-year-old married Kashmiri tribal woman $G_2P_2A_0L_0$ presented to the emergency department with severe weakness, bilateral flank pain and few episodes of non-bilious vomiting. Her medical history reveals that she was suffering from hypothyroidism. An examination of her nervous system revealed that there was proximal muscle weakness (MRC scale 4/5) and that her reflexes were flaccid. There was no evident muscle tenderness or sensory deficit.

Laboratory tests revealed she had deranged KFT with Urea BUN -71.30 mg/dl, Creatinine – 2.14mg/dl with an estimated Glomerular filtration rate [eGFR] of 29.3ml/min/1.73 m^2, mild hypokalemia K^+- 3.10, normal anion gap metabolic acidosis with Ph-7.26, HCO_3-8.2 mmol/L, PCO_2-18.6mmHg, with mild alkaline urine Ph-7.5. Her Complete blood counts revealed normocytic normochromic anemia (Hb-9.7g/dl, MCV-80Fl, MCH-28.1pg) with leukocytosis (WBC- 17.3K with 85% neutrophils). Her LFT showed elevated bilirubin (2.91mg/dl) that subsequently improved and dropped to 1.01mg/dl, ALP dropped to 127U/L from initial 172 U/L. Her serum calcium (9.9mg/dl) & serum PTH (57.6) were normal, ESR-110mm/hr., CRP was reactive, ANA was significantly positive (108.71).

Ultrasonography of her abdomen and pelvis revealed bilateral medullary nephrocalcinosis along with left grade I hydronephrosis secondary to left upper ureteric calculus. She also reported of having dry mouth and dry eyes however Schimer's test (OD-5mm, OS-5 mm), a short tear breakup time was >10 seconds and were normal. The American European Consensus Group (AECG) and the American College of Rheumatology (ACR)/European Alliance of Associations for Rheumatology (EULAR) provide classification criteria that serve as a useful guide for diagnosing SS. In practice, a diagnosis by a clinician is usually less stringent than would be accomplished using the full AECG or ACR/EULAR criteria and includes clinical experience and judgment. As an example, early in the disease course patients may have preserved gland function that would not meet the established threshold of positivity (Jonsson et al., 2018).

Other laboratory tests revealed that normal rheumatoid factor (RF = 10 IU/mL), anti-ds DNA antibody was negative (156.60), anti-Ro/SSA was 107.94 i.e., strong positive and normal anti-La/SSB antibodies (15.16), C_3 & C_4 complement proteins were normal & antibodies to Smith antigen are negative. Urine sediment was normal. Serology for hepatitis B virus, hepatitis C virus, and human immunodeficiency viruses were negative. The patient underwent a parotid salivary gland biopsy which was negative for focal lymphocytic sialadenitis. According to the AECG guidelines, SS is diagnosed when four out of the six items are present, including salivary gland pathology or the presence of autoantibodies against SS-A/SS-B [5].

The presence of sicca symptoms, the positive anti-SSA antibodies and Schirmer test confirmed the diagnosis of primary Sjögren's syndrome (total score = 4/9), according to the most recent ACR/EULAR Classification Criteria.

All appropriate consent forms from the patient for the publication of the case report were obtained.

Discussion

dRTA is an uncommon extraglandular manifestation of primary Sjögren's syndrome. It is most likely due to an immune-mediated injury of the H^+-ATPase pump in the intercalated cells of the collecting tubules (responsible for distal proton secretion) that leads to loss of H^+-ATPase activity. dRTA can be classified as complete or incomplete; the former is characterized by metabolic acidosis with morning urine pH >5.5 and a positive urinary anion gap. The incomplete form presents with normal serum bicarbonate levels, but urinary pH fails to fall to <5.3 after ammonium chloride loading. The incomplete form is the most frequent however our patient presented with complete distal RTA.

Increased urinary K^+ wasting leading to hypokalemia is the most common electrolyte abnormality in dRTA. Severe undiagnosed hypokalemia may cause muscle weakness or even paralysis that may precede sicca symptoms for months or even years before the final diagnosis of primary Sjogren's syndrome, as in our case. Other dRTA manifestations include nephrolithiasis or osteomalacia (leading to bone fractures). Our patient presented with severe bilateral medullary nephrocalcinosis, which is quite a rare manifestation of renal tubular acidosis, especially when associated with primary Sjögren's syndrome. Immunogenetic studies had suggested that both diseases have a common genetic predisposition.

Thyroid dysfunction is frequent in primary Sjogren's syndrome patients and those prone to develop thyroid disorders are identified by thyroid-related autoantibodies or by rheumatoid factor and anti-Ro/SSA activity. Patients with primary Sjögren's syndrome have an increased tendency to develop other autoimmune diseases. Hypothyroidism was the most common autoimmune disease developed in primary Sjögren's syndrome patients during follow-up of 10.5 years and the said patient was also suffering from hypothyroidism. As our case il-

lustrates, in patients presenting with unexplained episodes of hypokalemia, muscle weakness, and nephrolithiasis or nephrocalcinosis the possibility of primary Sjögren's syndrome -associated dRTA should be explored further by appropriate medical history, physical examination, and laboratory testing. Diagnostic delays may lead to severe consequences that include lymphomas.

In conclusion, whenever we come across a case that was admitted in our department, where a patient with a preset diagnosis and on existing treatment plan presents repeatedly to ER with relatively similar complaints, consideration should be taken to escalate further investigations to overcome the confirmation bias which is one of the most common types of cognitive bias or decision-making bias. Confirmation bias is a tendency to seek, interpret and remember information that confirms existing beliefs (e.g., diagnosis), skewing perception and judgement. Recognizing this bias enables more balanced decision-making by fostering openness to diverse perspectives and evidence through critical thinking and engaging in opposite viewpoints. When kept in mind in medicine it significantly helps in reducing undue diagnostic delays and missing any potential life-threatening consequences.

Conflict of interest

There are no areas of conflict.

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