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# A Case of Renal Tubular Acidosis with Sjogren's Syndrome

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**Abstract:**

This case study presents an 18-year-old female with symptoms suggestive of renal tubular acidosis (RTA), ultimately diagnosed with Sjögren's syndrome. The case highlights the diagnostic challenges associated with overlapping symptoms of autoimmune disorders and renal pathologies, underscoring the importance of a comprehensive evaluation and interdisciplinary collaboration in elucidating complex medical conditions.

**Background:**

Renal tubular acidosis (RTA) is a rare renal disorder characterized by impaired acid secretion, resulting in metabolic acidosis. Sjögren's syndrome, an autoimmune disorder primarily affecting exocrine glands, can present with a spectrum of systemic manifestations, including renal involvement. The coexistence of RTA and Sjögren's syndrome poses diagnostic dilemmas due to overlapping clinical features and necessitates a thorough investigation to differentiate between primary renal disorders and autoimmune etiologies.

**Case Presentation:**

An 18-year-old female presented with complaints of abdominal and leg pain, along with easy fatigability. Physical examination revealed pallor, and laboratory investigations showed metabolic acidosis and electrolyte abnormalities consistent with renal involvement. Imaging studies confirmed medullary nephrocalcinosis, raising suspicion for RTA. Positive autoimmune markers, including anti-Ro and anti-La antibodies, prompted further evaluation for an underlying autoimmune etiology. Lip biopsy demonstrated lymphocytic infiltration and fibrosis, supporting the diagnosis of Sjögren's syndrome.

**Investigation Chart:**

| | |
|---|---|
| Urea – 32 - 24<br>Creatinine – 3.2 – 1.4<br>Sodium – 140 – 138 - 137<br>Potassium – 3.1 – 3.8 – 4.2<br>USG Abdomen:<br>Echogenic foci noted in B/L renal medulla.<br>P/O Medullary nephrocalcinosis.<br>ABG – Metabolic acidosis | Urine PH – 7<br>Urine Citrate – 10 mg/dl<br>Anti -RO & LA – Positive<br>Lip biopsy – lymphocytic infiltration and fibrosis |

**Discussion:**

The convergence of renal abnormalities suggestive of renal tubular acidosis (RTA) and positive autoimmune markers posed a diagnostic challenge in this case. RTA typically presents with acid-base disturbances, but the presence of autoimmune markers raised suspicion for an underlying autoimmune etiology, necessitating further investigation. The initial presentation with abdominal pain attributed to nephrocalcinosis prompted consideration of RTA, highlighting the importance of recognizing renal abnormalities as potential indicators of systemic disease.

The subsequent diagnosis of RTA, followed by the identification of positive autoimmune markers and lip biopsy findings consistent with Sjögren's syndrome, underscored the complexity of this case. The transition from

RTA to Sjögren's syndrome emphasizes the dynamic nature of autoimmune diseases and their ability to manifest with diverse clinical presentations. Importantly, the rarity of the co-occurrence of RTA and Sjögren's syndrome further complicates the diagnostic process, necessitating a meticulous diagnostic workup to elucidate the underlying pathology accurately.

Furthermore, the association between RTA and Sjögren's syndrome highlights the potential renal involvement in autoimmune diseases. Sjögren's syndrome, characterized by exocrine gland dysfunction, can affect multiple organ systems, including the kidneys, leading to varied clinical manifestations. This case serves as an exemplar of the intricate interplay between renal and autoimmune pathologies, emphasizing the need for a comprehensive diagnostic approach encompassing clinical, laboratory, and histopathological evaluations.

**Conclusion:**

In conclusion, this case underscores the diagnostic challenges associated with the coexistence of RTA and Sjögren's syndrome. The timely recognition of autoimmune markers and histopathological evidence facilitated the accurate diagnosis and appropriate management of the patient. This case emphasizes the importance of a comprehensive approach and interdisciplinary collaboration in navigating complex medical conditions involving renal and autoimmune pathologies.

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