Section: Biochemistry

Original Research Article

PRIMARY SJÖGREN'S SYNDROME PRESENTING AS ACUTE PANCREATITIS, DISTAL RENAL TUBULAR ACIDOSIS, AND CENTRAL NERVOUS SYSTEM INVOLVEMENT

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ABSTRACT

Sjögren's syndrome (SS) is a systemic autoimmune disease most commonly associated with dryness of the eyes and mouth, although involvement of other organs can also occur. Presentation with simultaneous multi-organ involvement at the onset is uncommon.

We describe a woman in her early 30s who presented with vomiting, abdominal pain, and generalized weakness. Laboratory evaluation showed acute kidney injury, acute pancreatitis, and severe metabolic acidosis. Further testing revealed distal renal tubular acidosis with persistently alkaline urine and hypokalemia. During hospitalization, she developed generalized seizures, and MRI of brain demonstrated symmetrical white matter abnormalities. Immunological workup showed strongly positive anti-SSA/Ro52 antibodies along with hypocomplementemia. After excluding infections, malignancy, and other autoimmune diseases, she was started on immunosuppressive therapy. Her clinical condition improved with recovery of renal function and resolution of seizures and pancreatitis.

This case highlights that primary Sjögren's syndrome may present with life-threatening multi-organ involvement even without sicca symptoms.

INTRODUCTION

Sjögren's Syndrome is the second most common systemic autoimmune rheumatic disease, which occurs in 0.5-1.0% of the population, predominantly in females by a ratio of 9:1.[1,2] Though traditionally considered to be characterized by sicca symptoms, a subset of patients, as high as 70% of primary Sjögren's Syndrome patients, develop extraglandular symptoms involving the kidneys, lungs, nervous system, and gastrointestinal system.[2,3] Among these, distal renal tubular acidosis is seen in 5-33% of patients with renal involvement and can precede sicca symptoms by a few years.[4,5] Whereas neurological involvement affects 2–25% of patients,[6,7] clinically apparent pancreatic disease is rare, despite subclinical evidence of exocrine pancreatic insufficiency in up to 37% of patients.[8,9]

The combination of acute pancreatitis, distal renal tubular acidosis, and central nervous system involvement as a presentation of primary Sjögren's Syndrome is a rare occurrence. This is a case of a young female who presented with a combination of these unusual findings, leading to the diagnosis of primary Sjögren's Syndrome, which was facilitated by the recognition of unusual biochemical abnormalities in the biochemistry laboratory.

CASE PRESENTATION

A woman in early 30's with no known comorbidities, presented to the emergency department of the Hospital, with a three day history of persistent vomiting, watery diarrhoea, diffuse abdominal pain and progressive weakness. She reported a recent hospitalization at an outside facility for identical symptoms, from which she had been discharged after resolution of her symptoms.

The patient did not complain of dry eye or mouth, joint pain, skin rashes, photosensitivity, Raynaud's phenomenon or any urinary symptoms. There was no history of alcohol consumption, tobacco use, or medication intake. Family history was unremarkable for autoimmune or renal disease.

On examination, she appeared acutely ill and dehydrated, with tachycardia (108 bpm), hypotension (96/62 mmHg), Kussmaul respirations (26/min) and diffuse epigastric tenderness with guarding. There was no parotid enlargement, lymphadenopathy, joint swelling or skin lesions. Neurological examination was unremarkable at admission.

Investigations

Hematology:

Persistent microcytic hypochromic anaemia (haemoglobin 8.0–10.0 g/dL, MCV 61.8–67.4 fL,

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MCH 19.8–21.6 pg) with an iron profile consistent with anaemia of chronic disease (serum iron declining from 98 to 29.8 µg/dL, TIBC low at 183–205 µg/dL, ferritin elevated at 334–347 ng/mL). Peripheral smear confirmed microcytic hypochromic morphology without schistocytes. ESR was 39 mm/hour.

Renal function:

Blood urea 254 mg/dL and serum creatinine 18.9 mg/dL on admission, improving progressively to 23 mg/dL [urea] and 0.8 mg/dL [creatinine] respectively by day 14 without dialysis (Table 1).

Electrolytes:

Persistent hypokalemia (K^+ 2.5–3.8 mEq/L) requiring ongoing supplementation, intermittent hyperchloremia (Cl^- up to 112 mEq/L), hypocalcemia (8.7 mg/dL), and hyperphosphatemia (7.9 mg/dL).

Acid-base status:

Severe metabolic acidosis on day 2 (pH 7.23, pCO_2 19 mmHg, HCO_3^- 8.0 mmol/L) with appropriate respiratory compensation. Transient improvement on day 5 morning (pH 7.40) was followed by recurrent acidosis with hyperlactatemia (pH 7.24, lactate 9.2 mmol/L) after seizure onset. Acid-base normalized by day 7 with overcorrection to mild metabolic alkalosis (pH 7.47, HCO_3 33.5 mmol/L) (Table 2).

Urinalysis and urine biochemistry:

Persistent glucosuria (4+) with normal serum glucose, albuminuria (1–2+), microscopic hematuria (10-25 RBCs/HPF), and urine protein/creatinine ratio of 1.33. Urine pH was persistently alkaline at 7.4 despite systemic academia & urine electrolytes: Na^+ 116, K^+ 14, Cl^- 132 mmol/L, confirming impaired distal urinary acidification consistent with Type 1 (distal) RTA.[10]

Pancreatic enzymes:

Amylase 240 U/L and lipase 419 U/L on admission, peaking at 350 and 790 U/L respectively on day 4, then declining (Table 3). CA 19-9 was 68 U/mL (normal <37 U/mL).

Inflammatory markers and coagulation:

CRP rose persistently (2.87 → 19.02 mg/dL). D-dimer >4,000 µg/L. PT/INR worsened from 16.1s/1.15 to 40.7s/2.9, with APTT rising from 34.7 to 55.6 seconds, before partially correcting by day 8.

Infectious work-up:

Blood and urine cultures negative.

Immunological work-up:

Anti-SSA/Ro52 antibodies strongly positive (+4)(Figure 1); complement C3 58 mg/dL (low), C4 12 mg/dL (low-normal); Rheumatoid factor Negative; Anti-TPO 3.06 IU/mL (negative); other ANA subtypes negative. Thyroid function was normal.

Liver function: SGOT rose from 14 to 63 U/L (likely reflecting non hepatic sources including seizure related muscle injury and pancreatic damage); SGPT remained normal (13–19 U/L). Bilirubin was normal throughout.

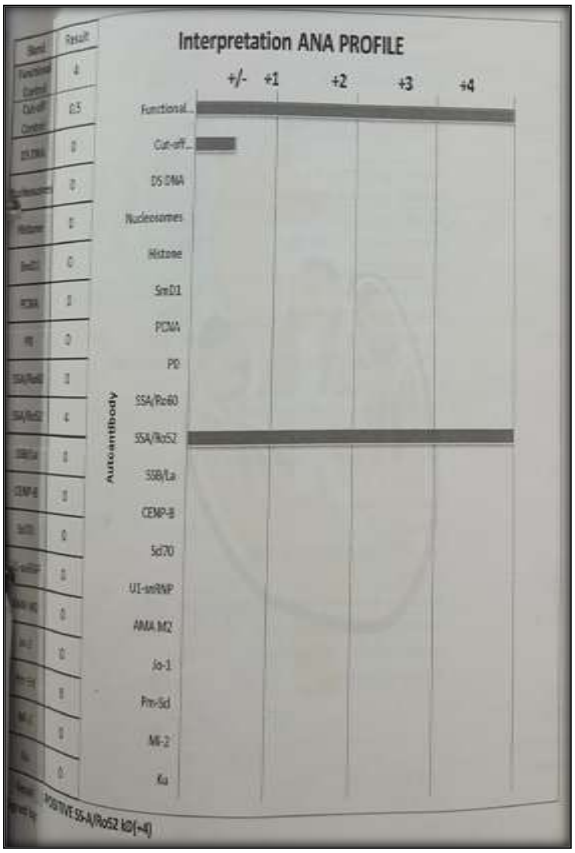

Figure 1: ANA Profile – Anti-SSA/Ro52 antibodies strongly positive (+4)

Table 1: Selected Serial Biochemistry Values

Parameter	Day 1	Day 3	Day 5	Day 7	Day 9	Day 11	Day 14	Day 15
Urea (mg/dl)	254	229	218	157	90	46	23	23
Creatinine (mg/dl)	18.9	16.4	10.2	5.6	2.2	1.4	0.8	0.8
K^+ (mEq/L)	3.2	3.7	3.3	3.1	3.2	3.0	3.9	3.6
Cl^- (mEq/L)	99	108	108	106	106	101	103	103
CRP (mg/dl)	2.87	7.14	8.42	10.11	14.98	-	19.02	11.26
Hb (g/dl)	10.0	8.8	-	8.7	8.7	9.2	9.0	8.8

Table 2: Arterial Blood Gas Trends

Day	pH	pCO_2 (mmHg)	HCO_3 (mmol/L)	Lactate (mmol/L)	Interpretation
Day 2	7.23	19	8.0	-	Severe Metabolic acidosis
Day 5 (AM)	7.40	35	21.7	-	Improved
Day 5 (PM, post seizure)	7.24	41	23.6	9.2	Recurrent Acidosis + Lactic acidosis
Day 7	7.47	36	33.5	-	Overcorrection to alkalosis

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Table 3: Pancreatic Enzyme Trends

Date	Amylase (U/L)	Lipase (U/L)
Day 2	240	419
Day 3	283	528
Day 4	350	790
Day 9	179	362
Day 15	182	442

Neuroimaging

On day 5, the patient developed two episodes of generalized tonic-clonic seizures. Non-contrast CT brain revealed hypodense areas in the midbrain and pons without haemorrhage, mass lesion, or midline shift.

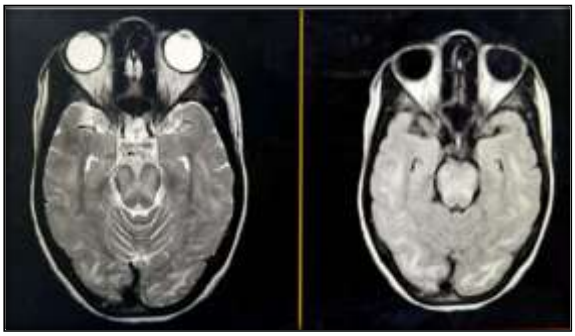

Figure 2: MRI Brain — Axial T2weighted (left) and FLAIR (right) images at the posterior fossa level demonstrating diffuse symmetrical hyperintense signal in the brainstem and bilateral cerebellar white matter.

Subsequent MRI brain (3D FLAIR protocol, without gadolinium due to renal impairment) demonstrated diffuse symmetrical T2/FLAIR hyperintense signal involving bilateral cerebral white matter (predominantly subcortical), bilateral cerebellar hemispheres, brainstem (pons), and splenium of the corpus callosum.(Figure 2) Patchy areas of restricted diffusion were noted in the bilateral posterior limbs of the internal capsules and occipitoparietal white matter. Ventricles, basal ganglia, thalami, and hippocampi were normal. The radiological impression favored toxic/metabolic encephalopathy with possible postictal changes, though the brainstem and cerebellar distribution raised additional concern for autoimmune or inflammatory pathology. [Figure 3]

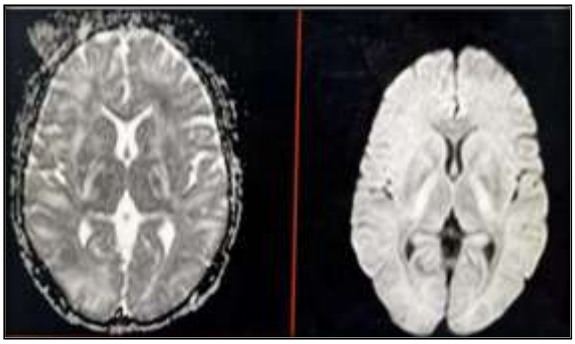

Figure 3: MRI Brain — ADC map (left) and DWI (right) showing patchy restricted diffusion in bilateral posterior limbs of internal capsules and occipitoparietal white matter

Abdominal Imaging

Ultrasonography demonstrated a bulky pancreas (head 3.5 cm, body 2.4 cm, tail 2.7 cm), bilateral raised renal cortical echogenicity with preserved corticomedullary differentiation, mild ascites, and echogenic mesentery with thickened bowel wall. No gallstones or renal calculi were identified.

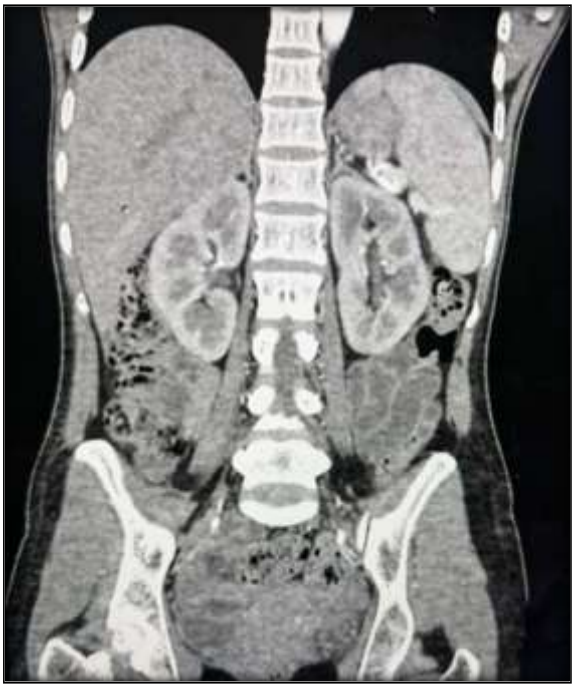

Figure 4: CECT abdomen (coronal reconstruction) demonstrating normal kidneys bilaterally, hepatomegaly, and normal pancreatic morphology without mass lesion or lymphadenopathy

Contrast-enhanced CT abdomen and pelvis showed hepatomegaly (16.3 cm) but a morphologically normal pancreas without any peripancreatic collections, mass lesions or ductal dilatation. No gallstones, lymphadenopathy or renal abnormalities were identified. Both kidneys appeared normal in size and density. Notably, the celiac trunk demonstrated a U-shaped stenotic configuration with poststenotic dilatation, consistent with median arcuate ligament syndrome (MALS); compression of the celiac axis by the median arcuate ligament of the diaphragm. [Figure 4]

DIFFERENTIAL DIAGNOSIS & DISCUSSION

This case presented an unusual diagnostic challenge: a young woman with simultaneous acute pancreatitis, dRTA with life threatening metabolic acidosis, and

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CNS involvement; none of which are typical presenting features of SS. The absence of sicca symptoms meant that autoimmune disease was not initially suspected. Instead, it was the recognition of biochemical anomalies that defied a single conventional explanation the paradox of alkaline urine during severe acidemia, normoglycemic glucosuria, pancreatic enzyme elevation without identifiable etiology, and anaemia of chronic disease in a previously "healthy" patient that prompted the immunological workup yielding the diagnostic anti-SSA/Ro52 result.

- **Renal Tubular Acidosis as a Harbinger of Sjögren's Syndrome**

The dRTA in SS results from auto-immune mediated destruction of alpha intercalated cells in the collecting duct, driven by lymphocytic infiltration and autoantibodies targeting carbonic anhydrase II and the vacuolar H^+ ATPase proton pump. [4,5,11] The classical biochemical triad hyperchloremic metabolic acidosis, hypokalemia and inappropriately alkaline urine was fully present in our patient. The concurrent normoglycemic glucosuria and proteinuria indicated additional proximal tubular dysfunction, consistent with the generalized tubule-interstitial nephritis described in SS.[4,5]

The bilateral raised renal cortical echogenicity on ultrasound, together with the anaemia of chronic disease at presentation, suggests subclinical autoimmune activity predating the acute presentation by a considerable duration, a finding consistent with reports that dRTA may precede sicca symptoms by years.[4,12] The rapid recovery of creatinine from 18.9 to 0.8 mg/dL without dialysis indicates that the severe azotemia was predominantly pre-renal (from volume depletion) superimposed on chronic tubule-interstitial injury.

- **Pancreatic Involvement**

Acute pancreatitis as a clinical manifestation of primary SS is rare, though subclinical exocrine pancreatic dysfunction has been documented in up to 37% of patients.[8,9] The pancreas shares embryological and functional homology with the salivary glands and is vulnerable to the same lymphocytic infiltration and immune complex deposition that destroys other exocrine tissues in SS [8]. In our patient, common etiologies (gallstones, alcohol, hypertriglyceridemia, drugs, and structural abnormalities) were systematically excluded through history, laboratory testing, and imaging. The mildly elevated CA 199 (68 U/mL) fell within the range expected for benign pancreatitis and should not be reflexively attributed to malignancy, particularly when cross-sectional imaging reveals no mass.[13,14]

An incidental but noteworthy finding was MALS on CECT. This anatomical variant, identified in 2–24% of the population, produces extrinsic celiac artery compression that can reduce pancreatic perfusion through the pancreatico-duodenal arcades.[22,23] While the majority of individuals with MALS remain asymptomatic, chronic subclinical pancreatic hypo-perfusion could theoretically render the organ more susceptible to acute injury from a superimposed autoimmune insult which is a potential “two-hit” mechanism. Additionally, the classical symptoms of symptomatic MALS (postprandial pain, nausea, vomiting) overlap considerably with this patient’s presenting complaints, raising the possibility that mild MALS related symptoms preceded and were masked by the acute autoimmune flare.

However, MALS alone cannot account for the dRTA, hypocomplementemia, anti-SSA/Ro52 positivity, or CNS involvement, and the clinical improvement with immunosuppressive therapy rather than vascular intervention supports autoimmune disease as the primary pathology. The degree of celiac stenosis was not quantified hemodynamically during the admission. If postprandial symptoms persist despite adequate immunosuppressive control, Doppler assessment of the celiac axis with respiratory maneuvers should be pursued, with consideration of surgical ligament release if hemodynamically significant compression is confirmed. [22,23]

- **Central Nervous System Involvement**

The neurological presentation in this case likely reflects dual pathology: metabolic encephalopathy from uremia, severe acidosis, hypokalemia, and hypocalcemia; all of which lower the seizure threshold,[15] acting synergistically with underlying autoimmune/inflammatory CNS disease. Several MRI features argue against a purely metabolic explanation: the prominent brainstem involvement (midbrain and pons), cerebellar white matter disease, splenial involvement, and restricted diffusion in the posterior limbs of the internal capsules, patterns that have been described in SS-associated CNS disease and are atypical for simple metabolic encephalopathy.[6,7,16]

Cerebrospinal fluid analysis was not performed, due to severe coagulopathy (INR 2.9) and clinical instability. Antiaquaporin4 and anti MOG antibodies were not tested. These investigations, along with gadolinium-enhanced MRI, are recommended during followup to further characterize the CNS involvement and exclude neuromyelitis optica spectrum disorder.[17]

- **Establishing the Diagnosis**

The strongly positive anti-SSA/Ro52 antibodies (+4), combined with hypocomplementemia (reflecting active immune-complex mediated complement consumption), anaemia of chronic disease, and multiorgan involvement typical of SS after systematic exclusion of SLE, IgG4related disease, sarcoidosis, malignancy, thrombotic microangiopathy, and posterior reversible encephalopathy syndrome supported the working diagnosis of primary SS.

Several key differentials merit specific mention. SLE was considered but argued against by the predominantly tubular (rather than glomerular) renal disease, negative ANA subtypes, absence of clinical criteria (malar rash, photosensitivity, arthritis,

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serositis), and pancreatic involvement [19]. IgG4related disease was considered because of its association with autoimmune pancreatitis and tubule-interstitial nephritis, but argued against by the demographic (typically elderly males), the acute presentation (rather than the painless obstructive jaundice typical of Type 1 autoimmune pancreatitis), normal pancreatic morphology on CECT, and anti-SSA/Ro52 positivity (not a feature of IgG4RD).[20]

It is acknowledged that the 2016 ACR/EULAR classification criteria for primary SS,[21] were not formally fulfilled during the acute admission: the anti-SSA/Ro positivity contributes 3 of the required 4 points, and objective sicca testing and labial salivary gland biopsy which could provide the additional point(s) were deferred due to clinical urgency and were recommended for follow-up.

- **Limitations**

Key limitations include the absence of objective sicca testing and labial salivary gland biopsy during the acute admission; lack of CSF analysis and serum IgG4 measurement; MRI without gadolinium contrast; absence of formal triglyceride measurement; and the patient's refusal of all recommended follow-up investigations, including confirmatory diagnostic testing and disease monitoring, resulting in incomplete characterization of the diagnosis and unknown long-term outcomes.

The degree of celiac axis compression in the identified MALS was not quantified, and Doppler assessment was not performed during admission.

Treatment

The patient was admitted to the medical intensive care unit and managed with aggressive supportive therapy targeting metabolic derangements, pancreatitis, and suspected infection.

Initial Supportive Management

Intravenous fluid resuscitation: Isotonic crystalloids (normal saline) were administered to correct dehydration and improve renal perfusion.

Correction of metabolic acidosis: IV sodium bicarbonate (8.4%) infusion was given due to severe metabolic acidosis (pH 7.23, HCO_3^- 8 mmol/L).

Electrolyte replacement: Persistent hypokalemia secondary to distal renal tubular acidosis was treated with oral potassium chloride syrup (Potklor) and IV KCl.

Antimicrobial Therapy

Empirical broad-spectrum antibiotics were initiated at admission due to systemic inflammatory response and concern for possible intra-abdominal infection:

Inj. Cefoperazone–Sulbactam 1.5g IV every 12 hours

Inj. Metronidazole 500 mg IV every 8 hours

Blood and urine cultures subsequently remained negative, and antibiotics were gradually de-escalated.

Symptomatic and Supportive Treatment

Tab. Hydroxychloroquine 200 mg/day

Inj. Ondansetron 4 mg IV every 8 hours for persistent vomiting.

Inj. Ranitidine 50 mg IV every 8 hours for gastric acid suppression

Inj. Tramadol (50 mg diluted in IV infusion) for analgesia related to acute pancreatitis

Inj. Vitamin K was administered for coagulopathy

Tab. Levetiracetam 500 mg every 12 hours.

Bowel rest and gradual reintroduction of oral feeding once symptoms improved

Hematologic and Nutritional Support

Tab. Mixzeal-XT once daily (iron, folic acid, and zinc preparation) was administered to address anemia.

Immunosuppressive Therapy

Inj. Methylprednisolone 1g/day for 3 days

Prednisolone taper: Started at 0.5/kg/day after pulse and tapered gradually.

Following identification of strongly positive anti-SSA/Ro52 antibodies with hypocomplementemia, the patient was started on systemic corticosteroid therapy to control autoimmune inflammation involving the renal tubules, pancreas, and central nervous system. This intervention was temporally associated with clinical improvement.

Hospital Course and Outcome

The patient was managed in the medical ICU with aggressive fluid resuscitation, intravenous sodium bicarbonate for metabolic acidosis, potassium and calcium supplementation, anticonvulsants for seizure control, conservative management of pancreatitis (bowel rest, analgesics, antiemetics), empirical antibiotics (de-escalated after negative cultures), and vitamin K for coagulopathy. Upon confirmation of anti-SSA/Ro52 positivity and hypocomplementemia, immunosuppressant (corticosteroids) therapy was initiated, with temporal correlation to clinical improvement.

Over 15 days, renal function recovered completely without dialysis, acid-base status normalized, seizures did not reoccur and pancreatic enzymes trended downward (though not fully normalized), coagulopathy improved, and CRP began declining.

The patient was discharged on oral bicarbonate and Syp Potklor 2 tbs BD, anticonvulsants [Tab Levetiracetam 500 mg OD], iron supplementation, and oral immunosuppressive therapy [Tab Prednisolone 40 mg OD] along with Tab HCQ 200 mg BD, with plans for outpatient follow-up.

At her first outpatient review four weeks post-discharge, the patient reported marked symptomatic improvement with resolution of abdominal pain, vomiting, and weakness. She was tolerating oral medications well and had no further seizure episodes. Physical examination was unremarkable, with stable vital signs and no new neurological deficits. However, she declined all further investigations, including objective sicca testing, citing financial constraints. Despite counselling regarding the importance of these investigations for confirming the diagnosis and guiding long-term immunosuppressive therapy, she did not return for subsequent appointments and was lost to further follow-up.

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Learning Points

- Distal renal tubular acidosis with alkaline urine and hypokalemia in a young patient should raise suspicion of Sjögren's syndrome, even without sicca symptoms.
- Primary Sjögren's syndrome may rarely present with multi-organ involvement, including kidneys, pancreas, and central nervous system.
- Early recognition and immunosuppressive therapy can lead to significant clinical recovery.

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