Paralysis in a young woman

case report

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A&E workup

- JBF, female, **32y.o**., caucasian
- Anamnesis: Rapidly progressive **tetraparesis** (within hours, no trauma), palpitations, polydipsia, polyuria
- Personal history: **iron deficiency anemia** (menstrual losses)
- Physical exam: **BMI 15,** normal BP, dehydrated and **anaemic conjunctivae**, slightly **swollen parotid glands**, **Premature ventricular contractions**, **flaccid tetraparesis: lower limb grade 1 muscular strength**, **upper limb grade 3 strength**, **hyperreflexia,** no meningeal or focal signs

A&E workup
+Paralysis

CNS related

- Spinal cord infarction (ischemic/hemorragic)

  80% painful, tetraparesis, arreflexia, pathological plantar reflex, distal sensatory loss

- Vertebral fracture / luxation
- Spinal cord / epidural haemorrhage
- Extradural: metastasis; vertebral tumours (sarcomas, plasma cell tumours); lymphoma
- Intradural-extramedullary: meningiomas; nerve sheath tumours; metastasis
- Intramedullary: ependymomas; gliomas; metastasis; round cell tumours
- Degeneratie myelopathy

A&E workup
+Paralysis

PNS associated

- Guillian-barré syndrome (acute polyneuropathy)

Motor and sensory symptoms, distal to proximal progression, flu-like prodrome, arreflexia

A&E workup

- ECG:
- Hemo
- MCV
- Leuco
- Plate
- **ESR (**
- CRP (
- Gluco
- ALT (U
- AST (
- Bil T
- CK (U

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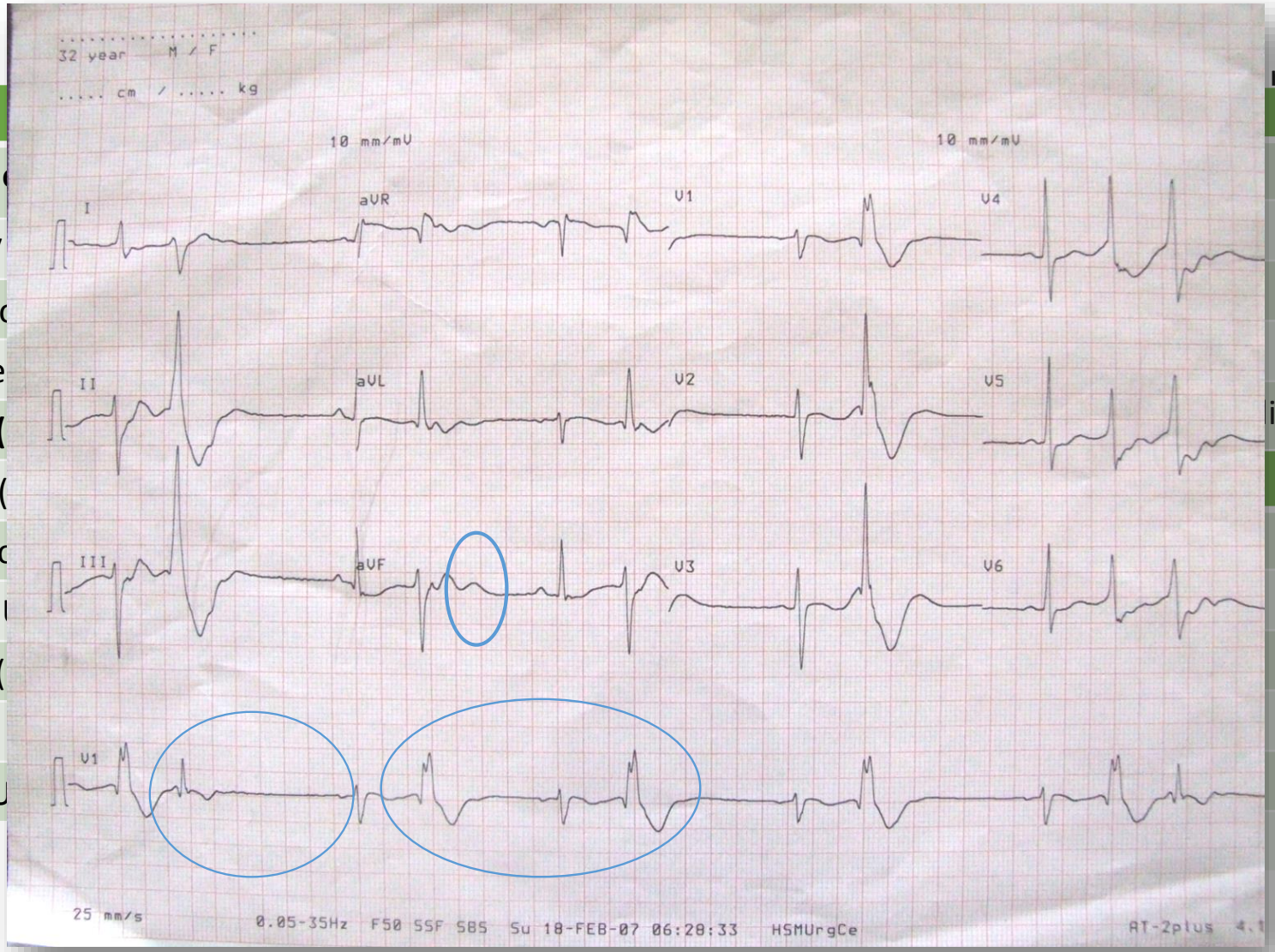


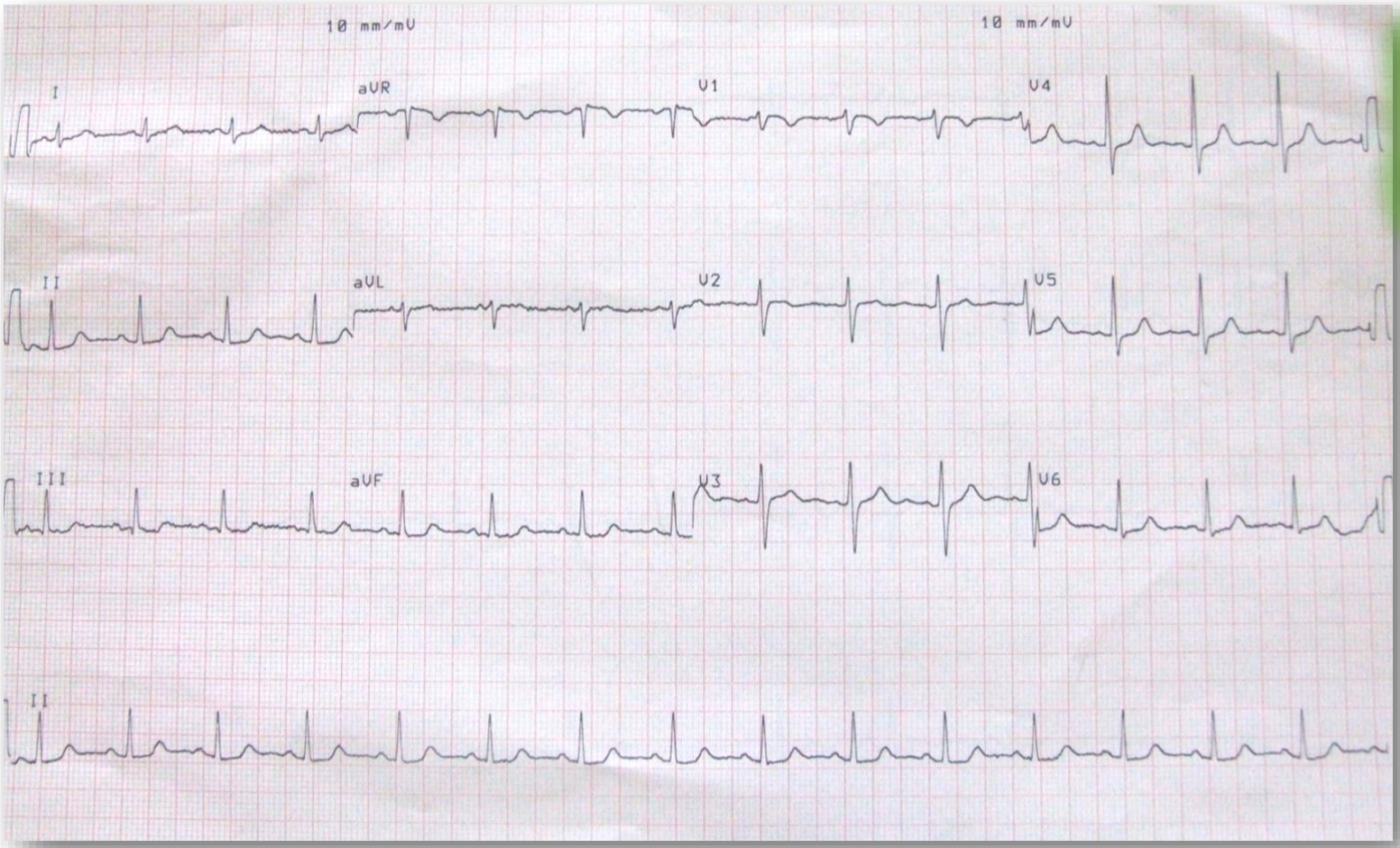
A&E workup
+Paralysis

Hypokalemia induced

- Nervous anorexia (intake, diuretics, laxatives?)
  - thiazide or loop diuretics: low BMI, hypokalemia, metabolic alkalosis
  - laxatives: hypokalemia, metabolic acidosis, no diarrhea
- Bulimia
  - hypokalemia, metabolic acidosis, low BMI, swollen glands, teeth pitting
- Tubular renal acidosis
  - Defective urinary acidification, hypokalemia, metabolic acidosis with normal AG, polyuria, polydipsia
- Hypokalemic periodic paralysis
  - stress or CH induced, hypokalemia, metabolic alkalosis

A&E workup
+Paralysis

Hypokalemia induced



Iv fluid and potassium correction was started, which partially reverted her neurological signs and normalized her ECG recording

Next diagnostic move?

Ward workup

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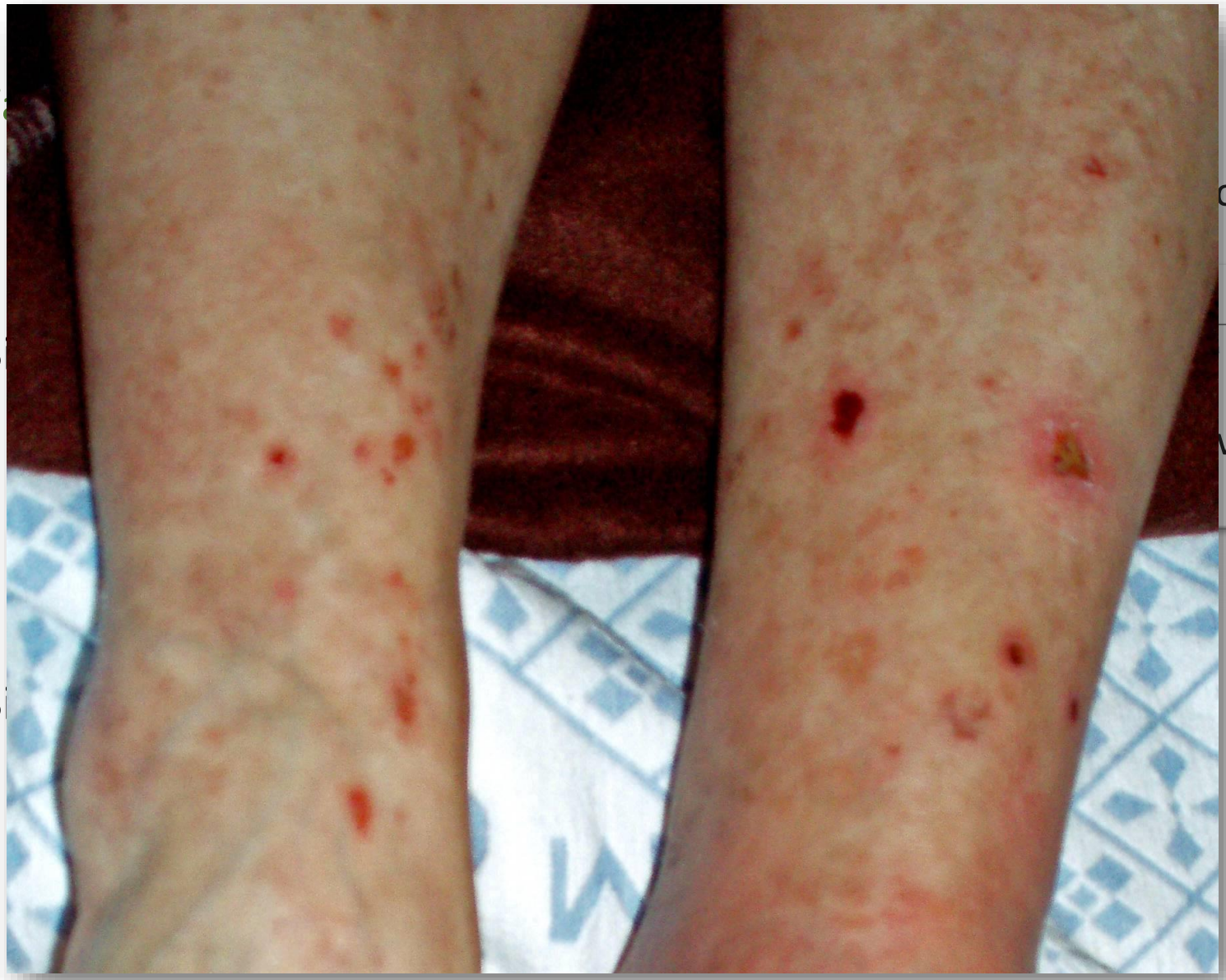
- S
- S



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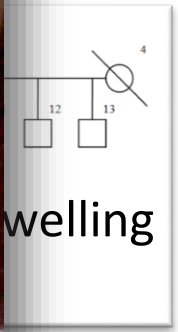
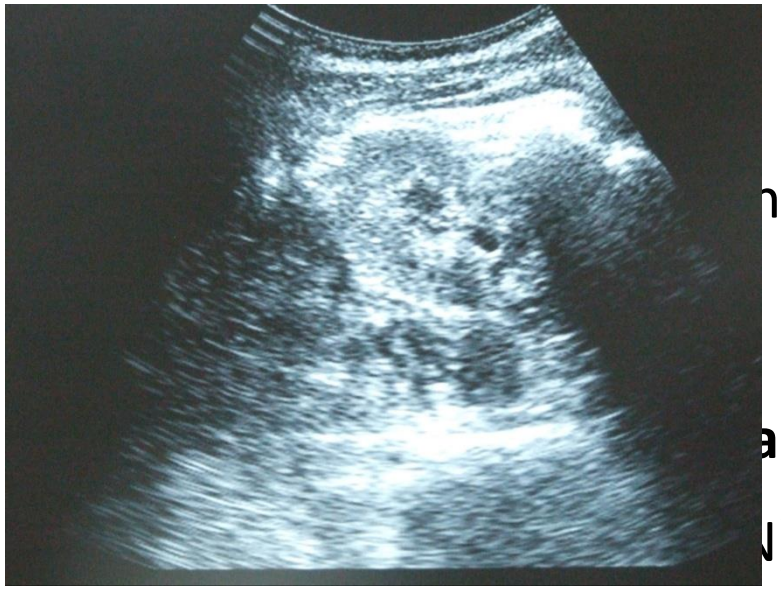
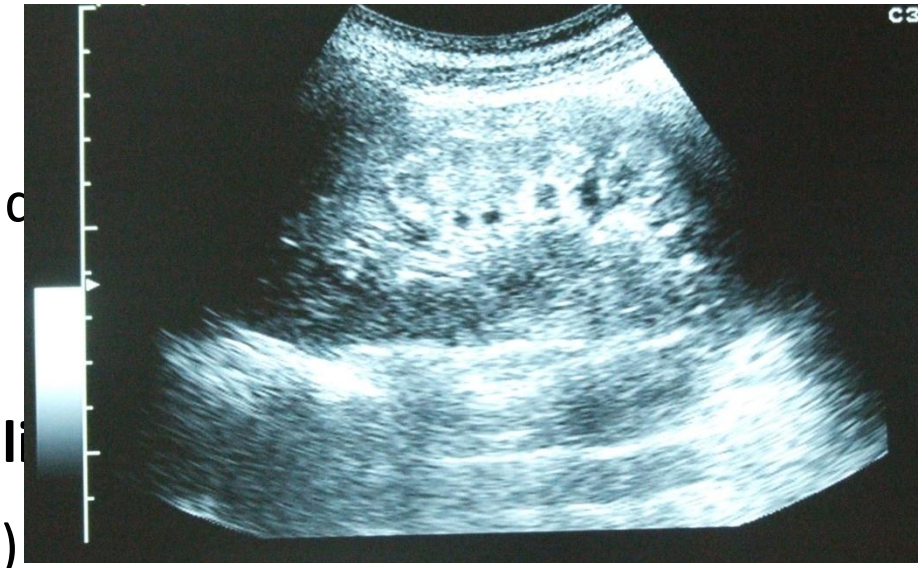


welling

Ward workup

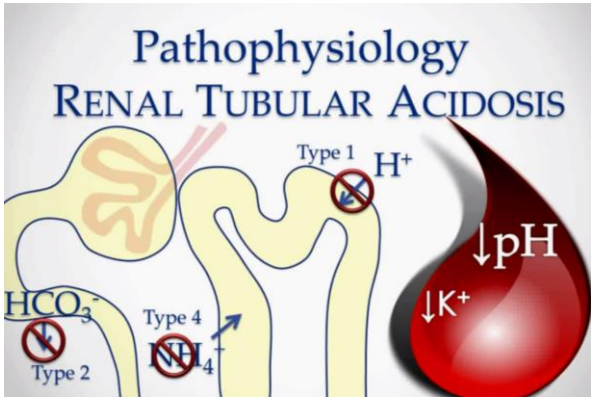
Renal disease





- **Urinary Phosphate (↑), calcium (↑), magnesium (↓), sodium (↑), potassium (↑), osmolality (↑),** Proteins (N)
- **Metabolic acidosis hyperchloremic**

Ward workup



Renal disease

- **Negative** Urine ammonium chloride test
- Urinary Anion Gap pos **19 mmol/L ($Na^+ + K^+ - Cl^-$)**

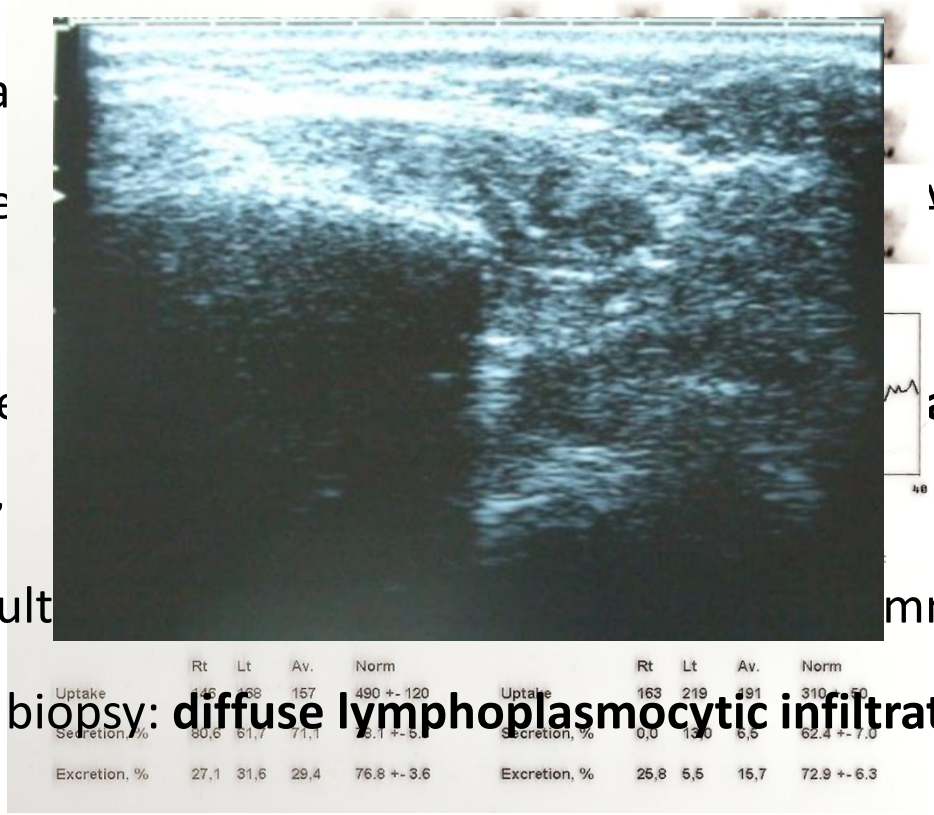
	V0	V1	V2	V3	V4
Interval(h)	0,5	1	2	2	2
Volume(mL)	40	40	700	240	200
pH (urine)	**8,07**	**7,81**	**7,32**	**7,65**	**7,33**
urinary pH	>5.5	<5.5	<5.5	5-6	
Amonium (urine)	125	125,7	117,1	128,9	133,8
H+ secreted (mmol/L)	Low	Normal	Low	Raised	
% HCO_3^- secreted	>10	>15	<10	<10	
Na^+ (urine) (mmol/L)	52	48	35	51	50
K^+ serum	Low	Low	Raised	Low	
K^+ (urine) (mmol/L)	46	51	23,7	32,3	40,6
urinary AG	Pos	Normal	Pos	Neg	
Cl (urine) (mmol/L)	71	75	33	62	83
pH (blood)	7,328	7,258		7,311	7,313
Sínd. Fanconi	No	Yes	No		No
Nefrocalcinosis	Yes	No	No		No

TRA 1 | TRA 2 | TRA 4 | GI losses HCO_3^-

Ward workup

Sicca complex

- **Positive anti-Ro/SSA and anti-La/SSB antibodies**
- **ANA 1:320** gra
- Negative RA te Ab, anti-centromerase Ab
- Parotid gland e **ands severe hypofunction**,
- Parotid gland ult mmetric glands
- Salivary gland biopsy: **diffuse lymphoplasmocytic infiltration,** normal B2 Microglobulin
- Schirmer test: RE 12 mm/5min, LE 15 mm/5 min
- Rose-Bengal test: no queratoconjuntivitis



Ward workup

Vasculitis

- Total serum proteins (N 80g/L)
- **Hypergammaglobulinemia** (18,6g/L), with **IgG** 19,90 g/L and **l** 2,85 g/L immunofixation

Anemia

- **Serum iron** 14 ug/dL (↓), **ferritin 35,3 ng/mL (↓)**
- Coombs test negative; normal LDH, haptoglobin and bilirrubin

Diagnosis

Primary Sjögren syndrome

- Type 1 tubular renal acidosis
  - Hereditary?
- Hypokalemic paralysis
- Mild chronic kidney disease
- Nephrocalcinosis
- Hypergammaglobulinemic purpura

Anemia ferropenic

Menstrual losses?

Diagnosis

SS Criteria	Definition
I. Ocular symptoms	- **dry eyes for more than 3 months** - recurrent sensation of sand or gravel in the eyes - tear substitutes more than 3 times a day
II. Oral symptoms	- **feeling of dry mouth for more than 3 months** - **persistently or relapssng swollen salivary glands** - **frequently drink liquids to aid in swallowing dry food**
III. Ocular signs	- Schirmer's test, performed without anaesthesia <(5 mm in 5 minutes) - Rose bengal score or other ocular dye score (4 according to van Bijsterveld's scoring system)
IV. Histopathology	**- Salivary gland biopsy**
V. Salivary gland involvement	- **salivary scintigraphy (schall score >1)** - parotide sialography - Sialometrics (≤1.5mL/15min)
VI. Autoantibodies	- **Antibodies to Ro(SSA) or La(SSB) antigens, or both**

American-European classification system

Therapeutics & Evolution

Therapeutics

- Potassium Chloride + Potassium citrate
- Monopotassic monophosphate
- Sodium bicarbonate (only after potassium resolution)
- Corticotherapy + Azathioprin
- Artificial tear + oral hydration ad libidum
- Oral iron supplementation

Evolution

- Full muscular strength recovery + cutaneous purpuric regression
- Hipokalemia+acidosis resolution
- BMI upgrade
- symptomatic relief

Discussion

- Sjögren's extra-glandular involvement
  - Arthralgy - 60%
  - Raynaud - 37%
  - Limphadenopathy – 14%
  - Pulmonary – 14%
  - **Vasculitis – 11%**
  - **Renal involment – 9%**
  - Hepatic involvement – 6%
  - Peripheral neuropathy – 2%

- Renal involvement in SS (Interstitial nephritis*)
  - Tubular renal acidosis type 1
  - Nephrogenic insipidus diabetes
  - Nephrolitiasis
  - Glomerulonephritis (rarer)

*The renal tubular acidosis in the presence of SS is usually due to **auto-immune lymphocytic tubular infiltration**

Take home message

Hypokalemia as a cause of **paralysis**

SS can lead to **cutaneous involvement** (hypergammaglobulinemic vasculitis); and also **kidney SS-involvement** characterized by renal tubular acidosis (absence of urinary acidification following the short ammonium chloride test)

The renal tubular acidosis in the presence of SS is usually due to **auto-immune lymphocytic tubular infiltration**

Severe hypokalemia, nephrolithiasis, nephrocalcinosis, chronic renal failure are among SS associated TRA1. These can be avoided if there is a prompt diagnosis leading to potassium supplementation and alkali replacement therapy.

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