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Photosensitivity for diagnosis

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Chief complaint

- 37-year-old European woman presented in P.Stradins university hospital with chief complains of cutaneous itching lesions and sensation of burning, skin fragility and blistering;
- Skin damage is confined to sun-exposed areas: face, neck, hands, forearms;
- Symptoms persist over 2-month period and clinical signs tend to become worse and wider;

History

2 month before the first consultation



4 day therapy with Sulfametaxazol/Trimetoprim 960 mg/ daily

2 days after the end of antibiotic therapy



itching small red cutaneous lesion in the left temporal region, that had transmuted into bulla, that made an erosion after splitting.

4 days after the end of antibiotic therapy



Diagnosis: Allergic dermatitis caused by Sulfametaxazol/Trimetoprim

First consultation



**P. Stradins
Clinical
University
Hospital**

Additional history

Anamnesis morborum:

- right renal aplasia;
- chronic kidney disease stage V;
- peritoneal dialysis (from Oct. 2011);
- secondary hyperparathyroidism;
- duodenal ulcer in remission;
- arterial hypertension;
- **Darbepoetin alfa** 30mkg weekly,
- **Calcitriol** 0,5 mcg 3x/week, **Sevelamer hydrochloride** 800mg 2x/day,
- **Perindopril/Indapamide** 2,5/0,625 mg daily,
- **Omeprazol** 20mg/daily, **Fucidin cream** 2% locally (arms, face).
- No similar complains in family members.

General examination



- Skin fragility
- Blistering
- Erosions

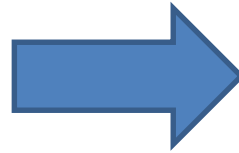
- Scarring
- Hypertrichosis
- Scarring alopecia



- Hyperpigmentation
- Hypopigmentation
- secondary infection

Workup....

Just collected urine



After 2 hours



Macroscopic fresh random urine analysis shows colour change within 2 hours.

Workup

✓ Liver impairment

- **ASAT – 67 U/I** (R<31), **ALAT 160 U/I** (R<31), **GGT – 55 U/I** (R<36),
- albumin, bilirubin – within the normal range;
- vaccinated against HBV; antiHCV – negative.

✓ Chronic kidney disease with PD, secondary hyperparathyroidism

- **creatinine 537 $\mu\text{mol/L}$** (R 30-80),
- **urea 15,5 mmol/L** (R 1,7-8,3),
- K 3,2 mmol/L (R 3,5- 5,3), Na 138 mmol/L (R 135-150),
- **Ca 2,45 mmol/L** (R 2,5-7,5), **P 1,37 mmol/L** (R 0,8-1,6), **PTH 20,2 mkmol/L** (R 1,2-5,8)

✓ Disturbance in iron metabolism and anaemia

- **HGB 10,3 g/dL** (R 120-153) (normocytic, normochromic)
- **ferritin – 1877 ng/ml** (R 12-122), iron – 6,1 mkmol/l (R 6,6-28,3), transferrin – 1,6 g/l (R 1,9- 3,3), transferrin saturation – 15% (R 12-45)

✓ Rheumatological tests within the normal range (ENA, ANA, ANCA, HIV Ag, RF).

✓ Urine test - **specific weight 1,009** (R 1,012-1,030), **urobilinogenuriya** (Bi- 17; R-neg.), otherwise – normal.



Porphyria cutanea tarda

.....endocrinologist, dermatologist consultation

..... skin biopsy

Workup

- ✓ Biochemical quantitative urine analysis (24-hour urine) showed:
 - ✓ high level of **coproporphyrin** 1098 mkg/g creatinine (R 20-80);
 - ✓ **delta aminolevulenic acid** (0,9 mkg/g creatinine) – within the normal range
- ✓ **Genetic analysis** as an **uroporphyrinogen decarboxylase (UROD)** enzyme activity assay in red blood cells - **are not available in Latvia.**
- ✓ **Skin biopsy** from affected skin region: dermal vascular superficial lichenoid dermatosis.

Therapeutic approach

- Sunlight avoidance (cream with SPF ≥ 50)
- Dietary recommendations:
 - reduce consume of meat (exspecially liver), butter, soybean. Alcohol restricted.
- «Save drug» list / «Prohibited drug» list
- Phlebotomy

Therapeutic approach

- **Renal transplantation** was made after 2 month of incident from deceased donor followed by standart 4 component immunosupresive therapy
 - basiliximab (monoclonal IL-2R antibodies) 2 injection on 0. and 4th day after transplantation;
 - glucocorticoids (metilprednisolon puls 3 days long, than continued with oral presnisolon 0,5 mg/kg/dn);
 - tacrolimus (calcineirin inhibitor) 0,1 mg/kg/dn with increasing dose to optimal (concentration in blood 5-10 ng/ml); to reduce the nephrotoxicity, the average concentration of tacrolimus in the blood kept at subtherapeutic levels during the first mounths after RT.
 - mucofenolate mofetil 1,5 g/dn;
- **Transplanted kidney biopsy** with histological examination of the obtained material: focal tubular necrosis without signs of acute transplant rejection.
- Additional therapy after transplantation included: anticoagulants, small doses of loop diuretics, broad-spectrum antibiotics, antifugal and antiviral medications.
- Discharged from the hospital after 18 days from renal transplantation.

- In outpatient treatment stage improved liver and transplant function, ferritin and coproporphyrin levels returned within the normal parameters.

Test	Before RN	18 days after RN	After half a year
Biochemical blood analysis			
Creatinine (mmol/L)	0.71	0.25	0.15
Urea (mmol/L)	14	13	9.1
Potassium (mmol/L)	4.4	4.4	4.5
Total protein (g/L)	63.7	54	64.4
Phosphorus (mmol/L)	1.39	1.2	0.8
Calcium (mmol/L)	2.50	2.4	2.6
ALAT (U/L)	88.5	52	13
ASAT (U/L)	58.8	42	13
Total cholesterol (mmol/L)	4.5	5.2	5.1
Glucosa (mmol/L)	5.4	5.0	4.8
Ferritin (ng/L)	1088	1021	253
Tacrolimus blood conc. (ng/mL)	-	2.5	6.7
Complete blood count			
Leu (cells/mL)	11.4	12.1	7.0
HGB (g/dL)	11.6	8.0	11.9
Plateles (mL)	327.000	237.000	176.000
Coproporphyrin in urine (mg/g creat)	-	156.0	41 (N)

9 days after renal transplantation



6 month after renal transplantation



skin ulcers were completely healed;
hyperpigmentation and hypertrichosis significantly narrowed;
Improved patient's mental health;

Thank you!

