Atypical cause of proteinuria – case presentation

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Young man (*1984)

- Admitted after injury with scrotal oedema
- Since puberty transient oedemas of scrotum, opalescent urine, continuus weight loss, no GI symptoms
- Nothing interesting from personal history
- 182cm, 58kg, BMI17,5 kg/m²
- 125/70 mm Hg, HR 78/min, BR 16/min, afebrile, asthenic, **slight scrotal oedema without any sign of inflammation,** otherwise normal physical finding

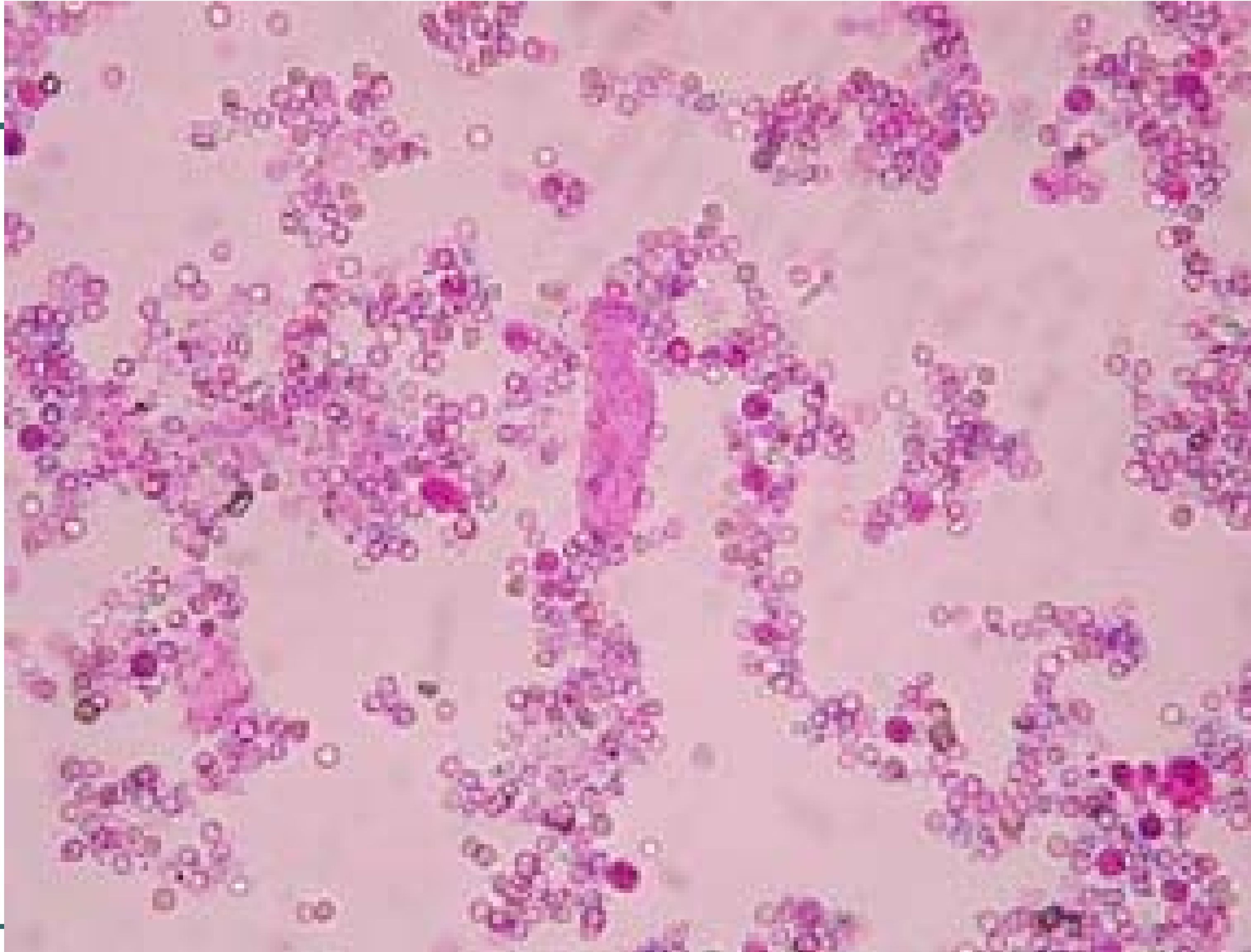
Lab test

- Na 133; K 5,1; Ca 2,4; P 1,3
- Urea 6,6 mmol/l; **S-crea 102 umol/l,** uric acid 365 umol/l
- **Cholesterol 2,9 mmol/l**, TAG 1,15 mmol/l
- **Albumin 24,4g/l**
- S-protein elfo: low albumin, low γ-globulin
- immunoglobulins: **low IgG, normal IgE,** normal IgM
- **FW 7/15, CRP< 3mg/l**
- Liver test: ALT 1,18 ukat/l, AST 0,99 ukat/l, otherwise normal
- **Normal blood count + differential**
- Normal thyroid, normal testosteron, normal cortisol

Urine analysis

- **Protein +3U**, otherwise chemically negative
- Ery 803/ul, leuko 17/ul
- **GF 1,74ml/s**, U-crea 6,2 mmol/l, BUN 281 mmol/day; TR 0,985; DU 2200 ml, **proteinuria 16,6g/day**
- U-protein elfo: mixed proteinuria
- U-immunofixation: no paraprotein

Ery phase contrast: extraglomerular erythrocyturia



Examination:

- Abdominal sonography: small ascites, kidney size and parenchyme normal
- MRI of scrotum: pathologic fluid in scrotum, punction: chylus
- CT of abdomen and small pelvis: free fluid otherwise normal
- CT enteroclysis: normal finding within small intestine

Summary:

- Severe proteinuria without nephrotic syndrome, extraglomerulal erythrocyturia
- hypoalbuminemia, low cholesterol
- Normotensive, normal US picture of kidney…. **What next?**
- **TAG in urine – 2,31 mmol/l**
- ➔ chyluria
- Kidney biopsy thus not indicated

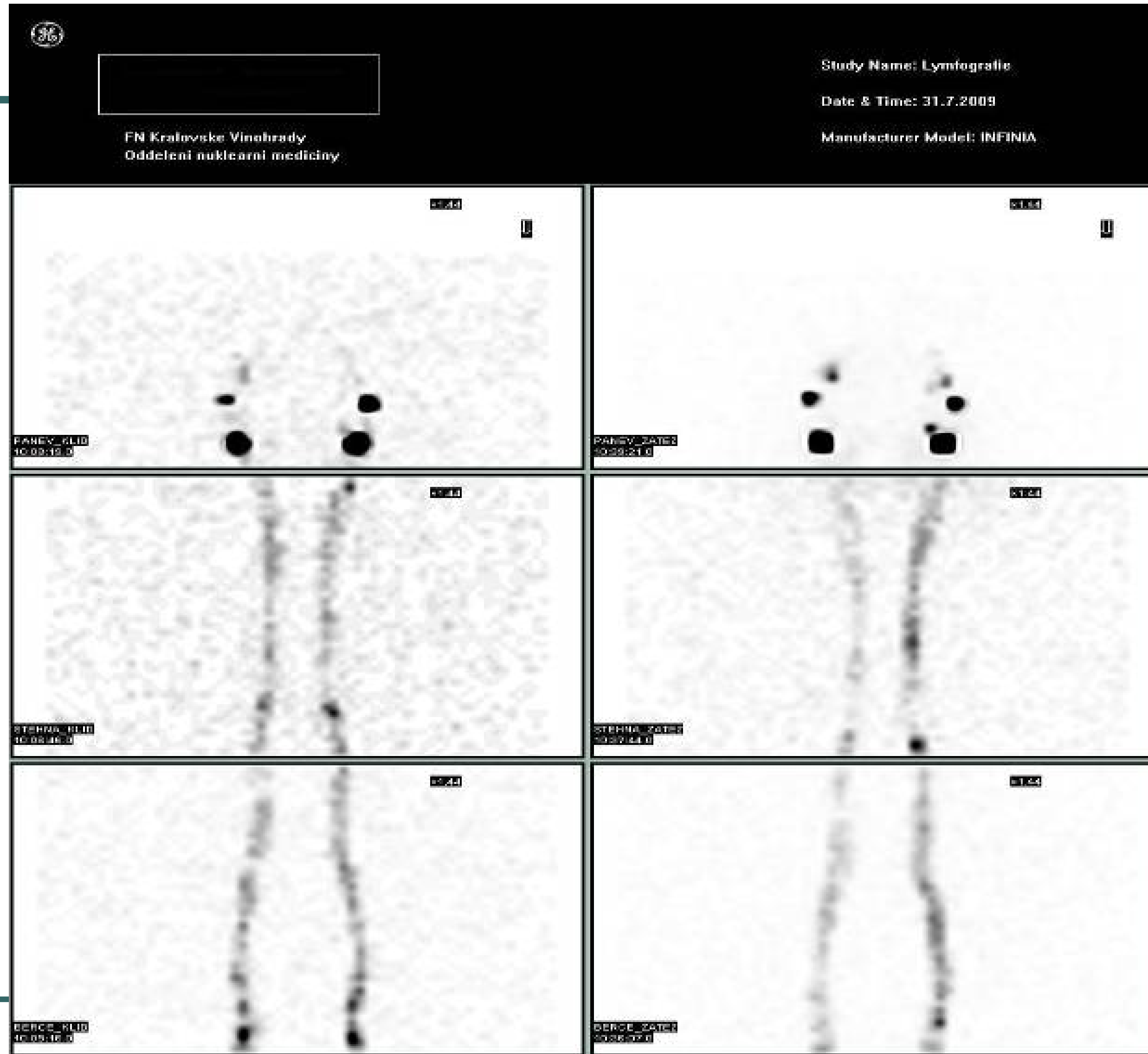
Background for diagnostic hypotheses:

- Chyluria with enormous urine loss of proteins
- Malnutrition marantic type
- Ascites, scrotal oedema – chylous

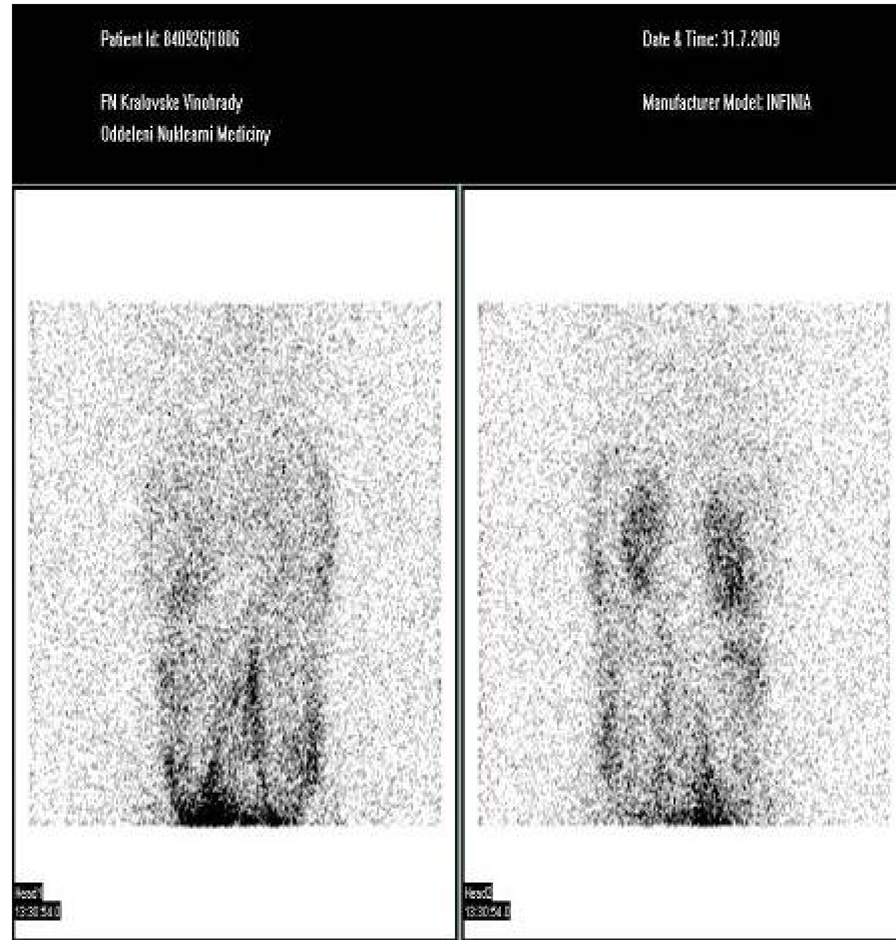
Dif dg

- disruption of intraabdominal lymphatics with pathologic communication between lymphatic and urinary system
- trauma, surgery
- infection (filariasis, tuberculosis, schistosomiasis, mesenteric adenitis)
- tumors
- lymphatic abnormalities (a/dysplasia, intestinal lymphangiectasias…)

Lymfoscintigraphy



Lymfoscintigraphy



Lymfoscintigraphy

- Borderline transport capacity of lymphatic system in lower limbs
- Pathologic accumulation of lympha in abdominal cavity
- Urinary lymphatic drenaige can not be excluded
- Susp. aplasia of lymphatics in abdominal cavity

Management

- Physiological background
- LCFA from gut via lymphatic system to venous circulation
- MCFA via vena portae to liver
- Decrease in alimentar LCFA could lower production of lympha and thus chyluria and oedemas, it will improve then nutritional status

Management and Outcome

- diet 2100kcal
- 105g of proteins
- no LCFA, only MCFA
- patatoes, pastry, rice, legumes, jam, honey, fruit, vegetables
- Nutridrink juice, Protifar, MCFA oil
- after 3 months: slow regression of oedemas, improvement of nutritional status

Conclusion

- chyluria can be confused with nephrotic syndrome
- chyluria occurs in disruption of lymphatics and pathologic communication (lymphatics-urinary tract) due to different causes (trauma, infections, dysplasia, surgery, tumor)
- not known possible treatment of aplasia of abdominal lymphatics
- diet revision with exclusion of fat (LCFA) and administration of MCFA can be effective