ΕΛΛΗΝΙΚΗ ΝΕΦΡΟΛΟΓΙΚΗ ΕΤΑΙΡΕΙΑ
HELLENIC SOCIETY OF NEPHROLOGY
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ΝΕΦΡΟΛΟΓΙΑΣ
19-21 ΙΟΥΝΙΟΥ 2024
ΜΕΓΑΡΟ ΔΙΕΘΝΕΣ ΣΥΝΕΔΡΙΑΚΟ ΚΕΝΤΡΟ - ΑΘΗΝΑ

Clinical Case Presentation

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Patient’s history

- A 19-year-old female patient presented to the outpatient clinic with periorbital oedema, which she noticed for days.
- On examination, there was mild peripheral edema and the patient was normotensive.

- Laboratory findings
  - Levels of serum creatinine and urea were normal (sCr=0.8mg/dl, sUrea=20mg/dl). She had low levels of serum albumin (2.9 g/dl).
  - The 24-hour urine sample showed 4.5g/24h of total protein.
  - Examination of the urine sediment did not reveal microscopic hematuria or casts.

Patient’s history

- Past Medical History: heterozygous b-thalassemia
- Family Medical history: father with hypertension
- Previous medications: sporadic use of NSAIDs

Course of Treatment

- **September 2021**
  - A kidney biopsy was performed that revealed podocytopathy, most likely **Minimal Change Disease**
  - Negative serology: (ANA, anti-dsDNA, anti-PLA2R, HBV, HCV, etc.)
  - Prescribed high dose methylprednisolone (40mg)

- **November 2021**
  - After 4 weeks of high dose of methylprednisolone there was no remission (24h urine protein: 2 g/24h, serum albumin: 2.9 mg/dl).
  - Cyclosporine was added

Course of Treatment

- **July 2022**
  - After 6 months of combined cyclosporine and methylprednisolone treatment only partial remission was achieved (24h urine protein=1.4 g).
  - Next step: Rituximab was administered (2 doses of 1g/15 days apart)

- **November 2022**
  - As the patient was steroid resistant and did not respond well to cyclosporine and rituximab (24h urine protein: 3.5 g/24h) a repeat kidney biopsy was decided.

Course of Treatment

- **December 2022**
  - Repeat kidney biopsy revealed **FSGS**.
  - Kidney biopsy report:
    - 12 glomeruli
    - Increased glomerular volume with segmental sclerotic lesions: one perihilar lesion, two lesions between the vascular and urinary pole, two lesions at the urinary pole (tip lesion like sclerosis)
    - Focal interstitial fibrosis (15%)
    - Immunofluorescence (3 glomeruli): IgM (+) fine granular staining in the mesangium and some glomerular capillaries
    - Electron microscopy: Diffuse foot process effacement, no dense deposits, no other structures
  - Genetic testing (Whole Exome Sequencing) for genes that corelate to nephrotic syndrome and kidney disease was negative.

- In the mean time methylprednisolone was re-administered

Course of Treatment

- **April 2023**
  - We initiated immunosuppressive therapy with cyclosporine again, as the patient remained nephrotic (low serum albumin:2.6 g/dl) with edema and proteinuria (2.4 g/24h).

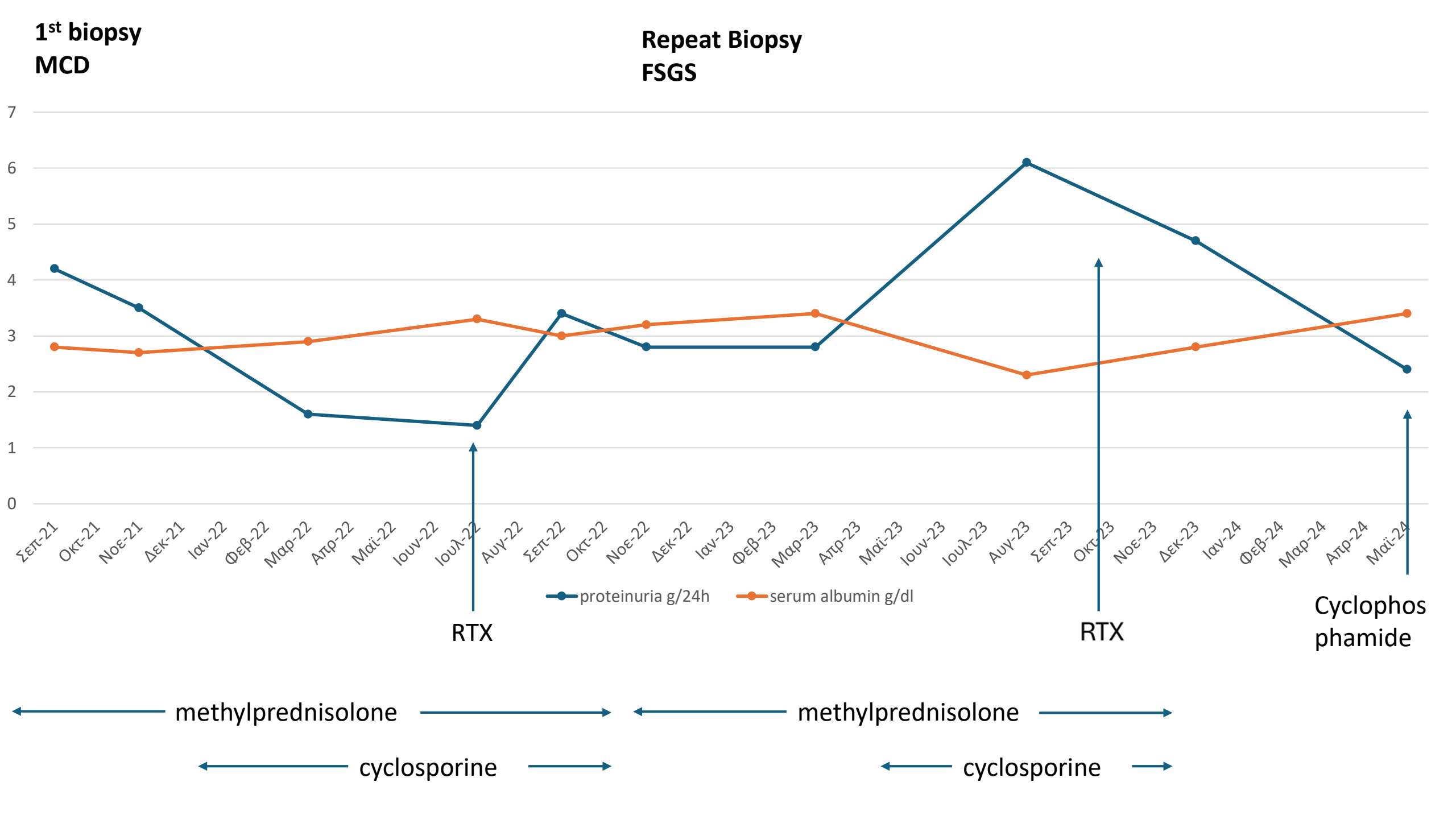
- **August 2023**
  - After 4 months of cyclosporine re-initiation the patient did not respond and reached her peak proteinuria levels at 6,1 g/24h.

- **September 2023**
  - A 2nd round of rituximab (2 doses of 1g, 2 weeks apart) was administered.

Course of Treatment

- **May 2024**
  - Upon persistent proteinuria (4 g/24h) and clinical features of sustained nephrotic syndrome Cyclophosphamide i.v. (750 mg) was administered. (at least 3 cycles of i.v. cyclophosphamide)
  - What is the next step?

- During the course of treatment, the patient also received maximum RAS blockade (irbesartan 300mg) which she tolerated well. In August 2023 she was prescribed Dapagliflozin as well. She also used diuretics (furosemide up to 120 mg/day) dependent on the state of the edema.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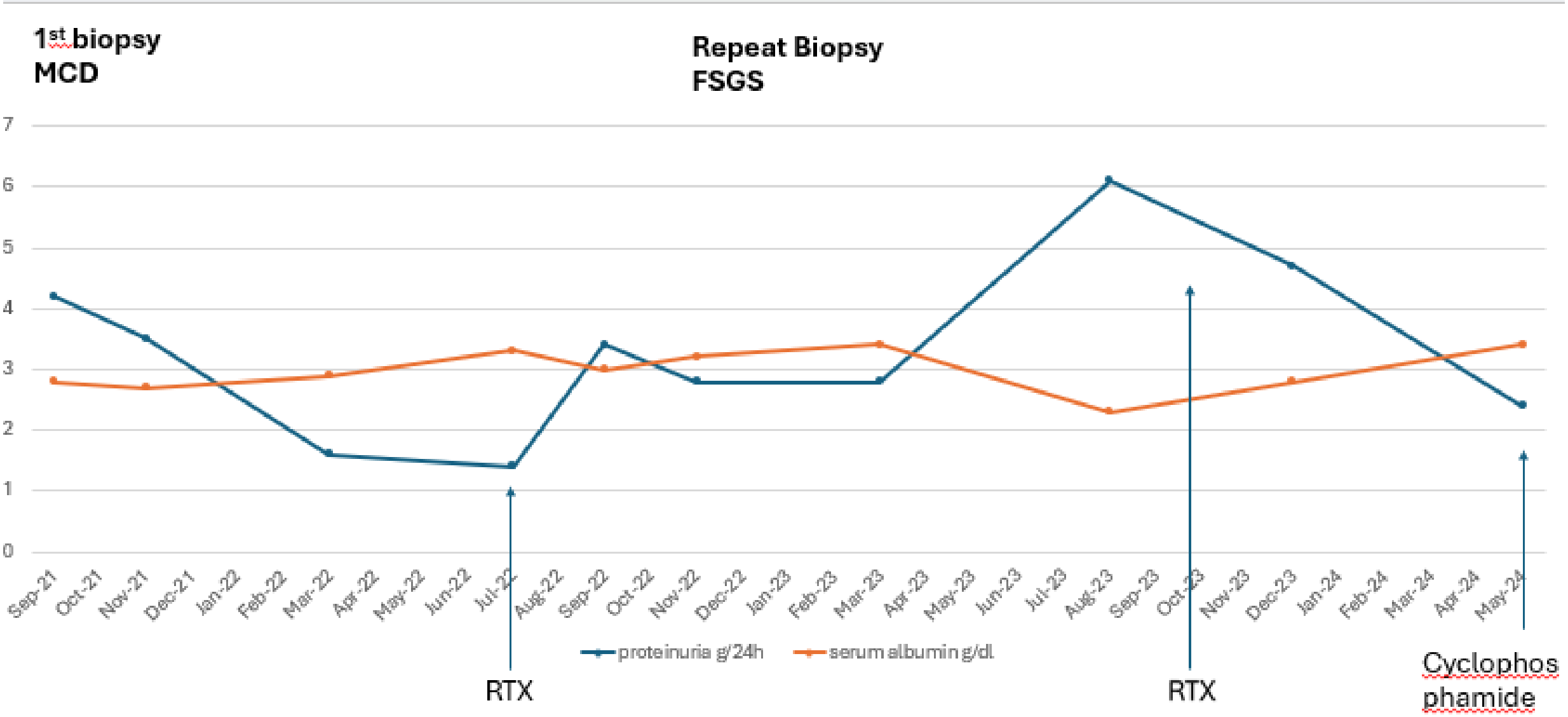
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