A DIAGNOSTIC CHALLENGE CASE REPORT: C3-GLOMERULONEPHRITIS IN A 17-YEAR-OLD ADOLESCENT

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INTRODUCTION

C3 glomerulonephritis is a rare immune-mediated disease caused by congenital or acquired disorders of the alternative pathway of the complement system, which debuts with the appearance of hematuria, proteinuria or acute kidney injury. The heterogeneity of the clinical picture determines the difficulty of diagnosing the disease. In particular, differentiate from clinically similar post-infectious glomerulonephritis (IRGN).

METHODS:

Clinical observation of a patient with hematuria, nephrotic-level proteinuria and an episode of acute kidney injury.

RESULTS

PATIENT M., A 17-YEAR-OLD ADOLESCENT

Acute respiratory viral infection

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Onset of disease

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At the time of admission:

- Creatinine increased to 368 µmol/l, dyslipidemia, total protein and albumin in the blood are normal;
- markers of systemic pathology are negative; C3 is transient decreased, C4 is normal, PLAR and IgG4 are normal; IgG is decreased; increase in D-dimer;
- eGFR CkiD U25 Cys - 46.2 ml/min/1.72m2;
- severe proteinuria up to 21 g/day persisted.

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IRGN was assumed, the onset of systemic lupus erythematosus and C3 glomerulonephritis was not excluded. Prednisolone 60 mg/day was prescribed.

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After initial of steroid therapy:

- Blood pressure: 154/88 mm Hg
- Creatinin increased to 137 µmol/l,
- Proteinuria remains up to 7,2 g/day;
- Due to the lack of effect, a puncture nephrobiopsy was performed

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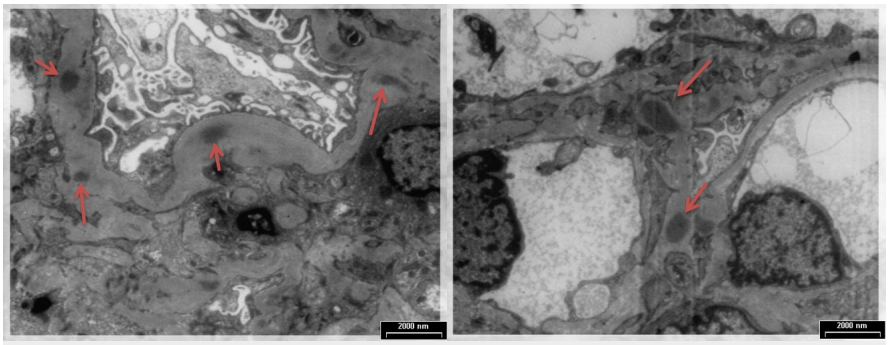

Figure 1. Intramembranous deposits detected by electron microscopy in a patient (authors' property).

Nephrobiopsy:
Pronounced mesangial proliferation, 53% of fibrous cell crescents, abundant deposition of IgG, IgA, and C3 in the mesangium and periphery of capillary loops, IgA, C1q - traces.
This morphological picture is most characteristic of C3 nephropathy.
According to electron microscopy - a picture of mesangioproliferative glomerulonephritis with deposits of various localizations: intramembranous, paramesangial and subpodocytic.

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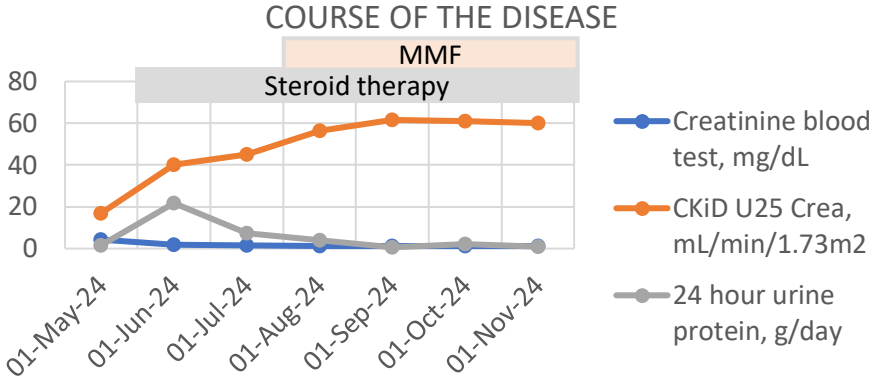

Figure 2. Patient's course of the disease

Taking into account the anamnesis data, nephrobiopsy and EM data, the adolescent has a picture of **C3 glomerulonephritis.**

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Immunosuppressive therapy (IST) was started - MMF at a dose of 900 mg / m2 and pulse therapy with methylprednisolone. Against the background of therapy - pronounced positive dynamics in the form of stabilization of renal function (eGFR CkiD U25 80 ml/min/1.72m2) and a decrease in proteinuria to 2 g/day.

CONCLUSION

Transient decrease in the C3 complement component can occur both in C3 glomerulonephritis and in acute post-infectious glomerulonephritis, which can significantly complicate diagnosis and increase the time before the start of therapy. In the absence of a clinical response to standard corticosteroid therapy, it is necessary to exclude C3 glomerulonephritis even with a normal C3 level by conducting a morphological study of the renal tissue. Pathognomonic for C3 glomerulonephritis is intramembranous deposition in the glomerular basement membrane by electron microscopy.