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Case Report:
Crescentic Glomerulonephritis in A Child With Tea-Colored Urine: A Case Report

Elham Vahhab[1], Mohammad Hossein Fallahzadeh[2], Anahita Dehghani[2], Babak Shirazi Yeganeh[3], Leila Malekmakan[2*]

1. Department of Pathology, Baylor University, Medical Center, Dallas, Texas, United States.
2. Shiraz Nephro-Urology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran.
3. Department of Pathology, Shiraz University of Medical Sciences, Shiraz, Iran

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Corresponding Author:
Leila Malekmakan
Address: Shiraz Nephro-Urology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran.
E-mail: malekmakan_l@yahoo.com

ABSTRACT

A 12-year-old girl was admitted with tea-colored urine for 24 hours. She had normal blood pressure and no edema with a rapid and progressive increase in blood urea nitrogen and serum creatinine, so one session of hemodialysis was done. The condition got worse by a thrombus formation in the heart. C4 was normal, but C3 was low, while serologic tests for lupus and Anti-Neutrophil Cytoplasmic Antibody (ANCA) were negative. Urinalysis showed proteinuria and hematuria. The kidney biopsy revealed crescentic glomerulonephritis. Treatment with 3 pulses of methylprednisolone and cyclophosphamide, and antithrombotic drugs were given with rapid recovery to normal serum creatinine and decrease in proteinuria. C3 was normalized after 2 months, and the clot disappeared, while hematuria and proteinuria continued as 1+ after 5 months. Due to the continuous proteinuria, Myfortic was continued for one year. Then, after stopping Myfortic, proteinuria did not return, and only microscopic hematuria continued. In conclusion, severe crescentic glomerulonephritis may have minimal symptoms and be complicated by heart thrombosis but with excellent response to medical therapy.

Keywords: Glomerulonephritis, Hematuria, Proteinuria, Biopsy

Introduction

Crescentic Glomerulonephritis (CGN) is usually the most severe form of glomerulonephritis that is uncommon in children [1]. It is defined as glomerulonephritis with the crescent formation in at least 50% of the glomeruli [2].

Rapid progressive GN can be classified as pauci-immune, immune complex-mediated, and anti-glomerular basement membrane diseases, while the pauci-immune type accounts for the most common variety [3]. Immune-complex mediated CGN causes severe glomerular damage and is associated with lupus nephritis, Henoch-Schonlein purpura, IgA nephropathy, and Post-streptococcal Glomerulonephritis (PSGN). The anti-glomerular basement membrane is an autoimmune disease that includes good pasture syndrome and isolated anti-GBM glomerulonephritis [4].

The prognosis and suitable management of CGN depend on the cause and the severity of the disease. But anti-GBM CGN has a poor renal prognosis that usually needs renal replacement therapy [2].

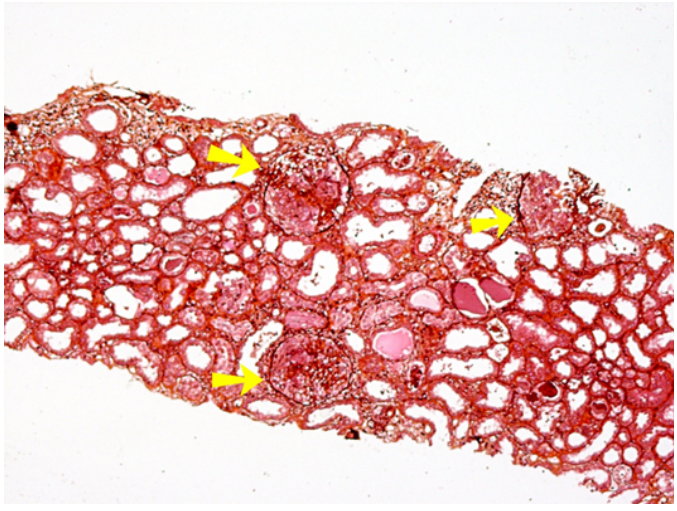

Figure 1. Kidney biopsy specimen showing 3 Glomeruli with cellular crescents, arrows (Jones Silver stain, original magnification 100x)

Here, we present a case of CGN with high serum Creatinine (Cr) level, normal Anti-Neutrophil Cytoplasmic Antibody (ANCA), and low C3 level with no definite cause.

Case Presentation

A 12-year-old girl, who was quite well up to 24 hours before admission, was presented with tea-colored urine. She had no history of edema or other symptoms except for oliguria and anorexia. On arrival, she had a good general condition with a blood pressure of 100/70 mm Hg and no other abnormal physical findings. Blood Urea Nitrogen (BUN) was 26 mg/dL, serum creatinine was 2.3 mg/dL that increased to 126 mg/dL and 7.6 mg/dL, respectively during 3 days.

Urinalysis revealed 1+ protein and 3+ blood while specific gravity was 1.011. Other paraclinical findings were as follows: cholesterol, 239 mg/dL; triglyceride, 268 mg/dL; hemoglobin, 13.8; serum albumin, 3.6 g/dL; ASO titer of 100. Also, ANCA and ANA and anti-double-stranded DNA were normal. C3 was low (48 mg/dL), while C4 was normal. Erythrocyte sedimentation rate decreased from 37 to 18, and proteinuria raised to 3+ while urine protein/creatinine ratio was 1.5, but still with no edema or hypertension. A double lumen catheter was

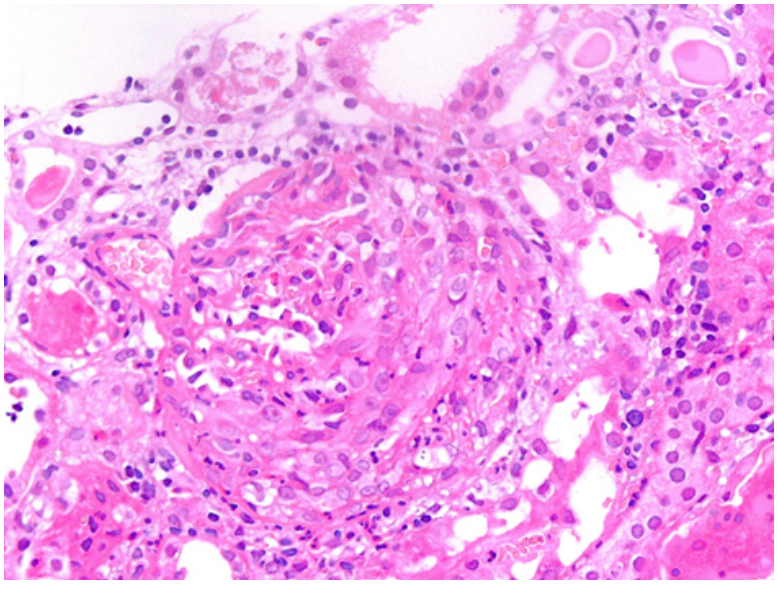

Figure 2. Glomerulus with cellular crescent, inflammatory cell infiltration and karyorrhexis (Indicating necrosis), (H&E stain, original magnification 400x)

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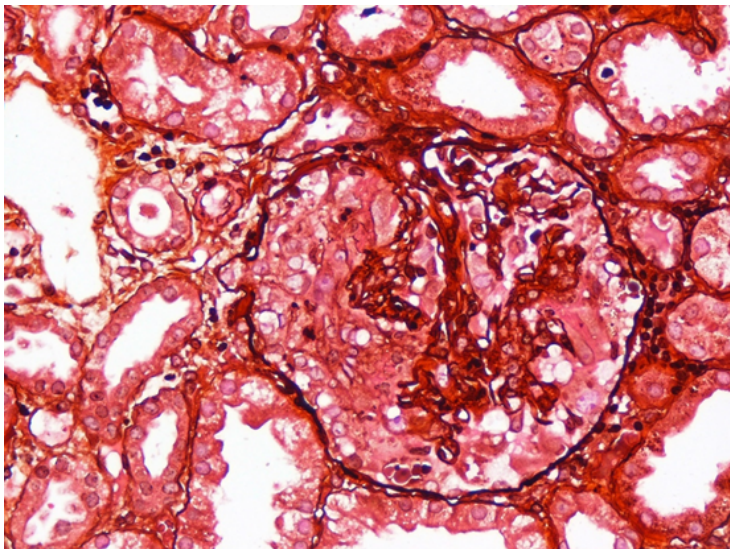

Figure 3. Glomerulus with cellular crescent, inflammatory cell infiltration (Jones Silver stain, original magnification 400x)

inserted for hemodialysis, and one session of hemodialysis was performed.

A kidney biopsy was done (Figures 1, 2, 3), and despite having no edema or hypertension, 3 pulses of methylprednisolone were given. After the report of CGN, one pulse of intravenous cyclophosphamide was also prescribed. In the biopsy specimen, there were 27 glomeruli; 2 of them were sclerosed, 21 of them with cellular crescent, and 4 with fibrinoid necrosis.

While the first echocardiography was normal, after hemodialysis, it revealed a clot with a diameter of 1.5 cm in the right ventricle. So heparin and dipyridamole were added to the prednisolone, and then heparin was replaced by warfarin. After four weeks, due to the persistence of proteinuria, Myfortic was also started while BUN became 10 mg/dL and serum creatinine decreased to 0.7 mg/dL. C3 was normalized after 2 months, and the clot disappeared while hematuria and proteinuria continued as 1+ after 5 months. Due to the continuous proteinuria, Myfortic was continued for one year, and proteinuria stopped after about 3 months of starting the Myfortic. By stopping Myfortic, proteinuria did not return, and only microscopic hematuria continued.

Discussion

This patient with CGN has had no clinicopathological correlation and was complicated by clot formation in the heart. CGN is uncommon in childhood and usually has a poor prognosis. It may be idiopathic or secondary to different diseases, including systemic lupus erythematosus, membranoproliferative glomerulonephritis, post-infectious glomerulonephritis, and vasculitis. Most of the childhood cases are pauci-immune with low C3. Almost all patients with Pauci-Immune Crescentic Glomerulonephritis (PICG) have circulating ANCA antibodies.

Diseases of CGN with low C3 are Systemic Lupus Erythematosus (SLE), Membranoproliferative Glomerulonephritis (MPGN), and PSGN. But in our patient with low C3, we have no evidence of PSGN by laboratory evaluations [5, 6]. Immunofluorescence on frozen tissue is the method of choice for diagnosing medical renal diseases; in our case, negative direct immunofluorescence of the renal tissue is against the group of immune GBM disease [7].

Consequences in CGN are disappointing, and progressive kidney failure has been seen in 18.9%-86% of patients. Better results may achieve by immunosuppressive therapy, cytotoxic drugs, and early dialysis or plasmapheresis. In our case, the presence of polymorph nuclear leukocytes may indicate an infectious process. Clot formation in the heart could be related to the double lumen catheter. Also, the drugs used for treating the clot may help control glomerulonephritis better. Anyway, despite diffuse crescent formation and proteinuria, the treatment protocol induced normalization of serum creatinine and disappearance of the clot and decreased proteinuria.

Rampelli et al. reported that renal survival of CGN depends on the age, etiology, extent of the renal failure, and the histological subtype [8]. Indian studies on CGN had reported End-Stage Renal Disease (ESRD) rates changing from 14.7% to 52.4%, but in contrast to our research, they had higher serum creatinine levels [9].

Patients with pauci-immune GN show a higher risk of ESRD. More studies are required to characterize the natural history of disease in children with ANCA-negative pauci-immune CGN; also, our patient's ANCA was negative with low C3 but with no definite cause [1].

Sadikoglu et al. reported a 6-year-old boy's case of CGN associated with infectious endocarditis with a serum creatinine level of 1.4 mg/dL that increased to 5.6 mg/dL and normal ANCA. Also, CGN with infective endocarditis has been reported in 11 adult patients by Kannan et al. [10, 11]; in contrast to this study, our patient developed a clot in the right ventricle that may be a complication of the dialysis catheter than the underlying disease.

Conclusion

The CGN has a wide range and includes various diseases, but our patient had no clinicopathologic correlation with no detected cause of CGN, and the final diagnosis requires multiple tests in her follow-up and consideration of the course of the disease in the future.

Ethical Considerations

Compliance with ethical guidelines

All ethical principles are considered in this article. The participants were informed of the purpose of the research and its implementation stages. They were also assured about the confidentiality of their information and were free to leave the study whenever they wished, and if desired, the research results would be available to them. A written consent has been obtained from the subjects. principles of the Helsinki Convention was also observed.

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Authors' contributions

All authors equally contributed to preparing this article.

Conflict of interest

The authors declared no conflict of interest.

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