

Images in Nephrology
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Smiling glomerulus reveals diagnosis in seronegative anti-glomerular basement membrane disease

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A 58-year-old woman with a history of tobacco use and chronic obstructive pulmonary disease was urgently referred by her family doctor for coffee-colored urine and swollen ankles since 2 days. The serum creatinine was 3.3 mg/dL, having increased from 0.9 mg/dL 1 month before. Blood pressure was 116/54 mmHg. The urine sediment showed acanthocytes and red cell casts and protein/creatinine ratio was 1.2. The chest X-ray showed no focal infiltrate. The patient was hospitalized and a renal biopsy was performed. Light microscopy revealed a focal necrotizing glomerulonephritis with crescent formation but only 2 of 14 glomeruli were sclerotic. Immunofluorescence showed linear staining for IgG in all seven evaluable glomeruli. There was a 'smiley'-like picture in one glomerulus (Figure 1). No deposits were found on electron microscopy. A diagnosis of anti-glomerular basement membrane disease (anti-GBM disease) was made and the patient was treated with steroids, cyclophosphamide and plasmapheresis. Testing for anti-GBM antibodies was negative in two different reference laboratories; thus, it would have been impossible to make the correct diagnosis without a biopsy. Published data indicates that serology is negative in up to 13% of cases of biopsy-proven anti-GBM disease [1].

The serum creatinine decreased from a peak of 4 mg/dL to baseline 0.8 mg/dL in 3 months. The patient quit smoking and after 6 months the cyclophosphamide was tapered. The patient had persistent microalbuminuria and microscopic hematuria. Two years later, the serum creatinine remained normal and her microalbuminuria had resolved, but there were still rare acanthocytes in the urine sediment. This case illustrates with a 'smile' that rapid referral and renal biopsy are crucial to achieving a good outcome in rapidly progressive glomerulonephritis.

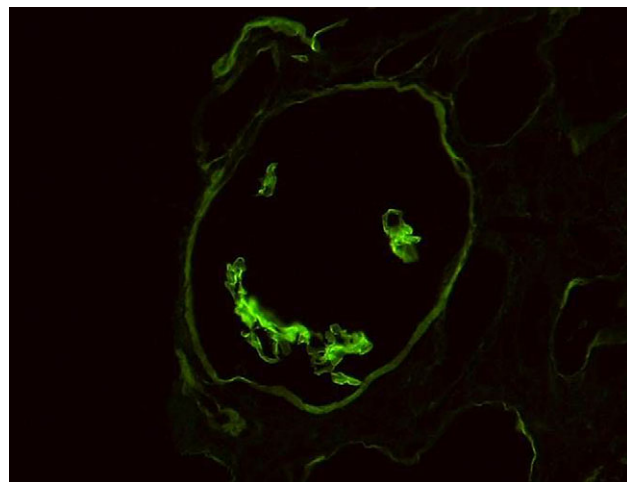


Fig. 1. Linear staining for IgG showing a 'smiley face'.

Conflict of interest statement. None declared.

References

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