Case Report

Nephrotic syndrome is a rare manifestation of IGA nephropathy

Ahmad A Alshomar

Medicine Department, College of Medicine, Qassim University, Kingdom of Saudi Arabia.

Abstract

Nephrotic syndrome is a rare presentation of IgA nephropathy. The degree of proteinuria in IgA nephropathy predicts poor prognosis. We herein report a teenager with IGA nephropathy, the nephrotic syndrome and segmental glomerular scars who after developing complications from high dose corticosteroid therapy was successfully treated with tacrolimus and low dose prednisone.

Key words: Nephrotic syndrome, IGA nephropathy

Correspondence:

Ahmad A Alshomar
Demonstrator,
Medicine Department,
College of Medicine,
Qassim University,
Kingdom of Saudi Arabia.
Mobile: +966566534423
Email: Ahmad.506@hotmail.com
Introduction

IgA nephropathy is the most common form of glomerulonephritis. (1, 2, 3, 4) Nephrotic syndrome occurs in 5-10% of IgA nephropathy, (5) although two studies from China report the frequency to be more than 30% in children. (6) Nephrotic range proteinuria in IgA Nephropathy was initially thought to represent an associated minimal change type podocyte injury with a favorable response to corticosteroid therapy. Subsequent reports indicated the presence of focal segmental sclerosis in association with IgAN. This has been confirmed as a negative prognostic factor. (7) Additionally, Proteinuria has been shown in multiple studies to be a predictor of outcome in IGA nephropathy. (8) With the rate of progression being very low among patients excreting less than 1g/day and greatest among those excreting more than 3 g/day. (9) Heavy proteinuria is generally considered to portend worse outcome. (10) In one study, the degree of proteinuria was found to be the most important predictor of the rate of GFR decline. In that study, the loss of renal function was 25-fold faster in those with sustained proteinuria ≥ 3 g/d than in those with < 1g/d. (8) This provides a strong rationale for the use of antiproteinuric interventions. We herein describe a teenager with IGA nephropathy and nephrotic syndrome who developed a complication of high dose steroid therapy but achieved a sustained complete remission with the use low dose combination of tacrolimus and low dose prednisone.

Case report

A 14-year-old Saudi male developed generalized edema and frothy urine. At the local hospital, he was found to be hypertensive with microscopic hematuria, normal creatinine and urine protein 10g/day. Renal biopsy was done and showed mild mesangial expansion on light microscopy. Immunofluorescence and electronic microscopy were not performed. A presumptive diagnosis of IGA nephropathy was made and the patient started on 60 mg per day of prednisone orally. High dose prednisone therapy was complicated by upper gastrointestinal bleeding requiring ICU admission. He was referred in our hospital for further evaluation and management. On Physical examination, he appeared malnourished. Blood pressure 126/77mmHg, pulse: 108 beats per minute and body mass index was 16. Cardiovascular, chest and abdomen were unremarkable. He has mild lower limb edema. There was no rash or joint swelling. labs; WBC:13.8, Hemoglobin:11g/dl, Platelet: 240, serum albumin:16 g/l, serum creatinine:48 micromole/l urea:4.9 sodium:136 K:3.6 total cholesterol :9.4mmol/l Triglyceride:3.2mmol/l, normal complement, Anti-nuclear antibody was negative, Anti-streptolysin O titer was unremarkable, Hepatitis B surface antigen and Hepatitis C virus antibody were negative. Urinalysis: protein (A+3), Blood (A3+), microscopy RBC (A 50/HPF), crystals, granular casts, RBC casts negative. A 24 hours urine collection of protein on admission 4.8g. Renal US was unremarkable. Renal biopsy was performed. Light microscopy revealed mild mesangial proliferation, no endocapillary proliferation, a few segmental scars and minimal interstitial fibrosis and tubular atrophy. Immunofluorescence was positive for IgA. Electronic microscopy revealed immune deposit confined to the mesangial with extensive foot process effacement (Figure 1). In view of patient GI symptoms, prednisone was kept at 20 mg daily and tacrolimus was added at 1 mg BID and the dose was adjusted to maintain a serum level of 3.5-5. On outpatient follow up, his proteinuria showed progressive decline. His prednisone dose was slowly tapered down. Seven months after the initiation of tacrolimus, his urine protein/creatinine ratio was 0.24, serum albumin normalized and his anemia resolved. The patient general condition improved.
Discussion

The Nephrotic syndrome occurs in less than 10 % of all patients with IgAN. The diagnosis requires a kidney biopsy. Clinical predictors of progression of IgA nephropathy include impaired renal function, hypertension, and persistent proteinuria above 1g/day. Renal survival was highest in patients who had no increase in proteinuria, followed by patients who had the smallest increase (ie, 1 to 2 g/day); renal survival was lowest among those with greater increases in proteinuria. Independent histological predictors include mesangial and endothelial proliferation, segmental sclerosis and tubular atrophy/interstitial fibrosis. Our patient had nephrotic proteinuria and segmental sclerosis as negative prognosticators. He was initially treated with high dose prednisone and developed upper gastrointestinal bleeding requiring ICU admission before having been referred to our center. We were reluctant to increase his corticosteroid dose in view of his history of GI bleeding and frailty. We elected to add low-dose tacrolimus aiming at achieving a remission of the nephrotic state. \(^{(1)}\)

Conclusion:

This case highlights poor prognostic features of IgA nephropathy. The use of low dose combination of tacrolimus and low dose prednisone may be consider as alternative treatment to high dose steroid because the side effect.

References:


Figure 1 showing (A) Segmental sclerosis-trichrome stain. (B) Segmental sclerosis-PAS stain. (C) Mesangial hypercellularity-PAS stain. (D) Mesangial proliferation –EM. (F) Mesangial deposited –EM.
