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Case Report

IgA NEPHROPATHY PRESENTING AS NEPHROTIC SYNDROME- AN UNUSUAL PRESENTATION

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ABSTRACT
Nephrotic syndrome is an unusual manifestation of IgA Nephropathy (IgAN). Some cases respond to steroid treatment. Here we describe a case of IgAN with steroid responsive nephropathy. The patient showed complete remission after steroid therapy. The clinical features of sudden onset generalized edema, gross hematuria, heavy proteinuria, initial hypoalbuminemia following a episode of upper respiratory tract infection might help identify the subset of patients.

INTRODUCTION
IgA nephropathy (IgAN) is the most common primary glomerulonephritis worldwide (D'Amico, 1987; Julian *et al.*, 1988). The dominant mesangial IgA deposits represent the diagnostic hallmark of IgAN (3,4). Gross haematuria bouts preceded by an upper respiratory infection, hypertension and proteinuria and microhaematuria of variable degrees are the most characteristic clinical findings. Dominant or co-dominant mesangial deposits of IgA, with IgG and C3, are the typical findings in renal biopsy immunofluorescence (Julian *et al.*, 1988). Its clinical features are highly variable. Although a moderate degree of proteinuria is common in patients with IgAN, nephrotic syndrome is considered uncommon in these patients. However, the response to steroid treatment has been variable, and usually correlated with the underlying histological changes.

CASE
A 19 year old male presented with the complaints of sudden onset one episode of gross hematuria, which was followed by swelling in b/l lower limbs which progressed rapidly to become generalized in two to three days. He also complained of sore throat and fever 3-4 days prior to the onset of hematuria. Clinical examination revealed normothermic individual with pulse rate of 86/min, blood pressure of 170/100 mm of Hg without any postural fall and respiratory rate was around 20/min. There was no pallor, icterus, lymphadenopathy, clubbing. Gross anasarca was present. Cardiovascular, chest and central nervous system examinations were within normal limits. Abdominal examination was s/o ascites, shifting dullness was present. Urine analysis revealed straw coloured, turbidity +2, pH- 6, specific gravity 1020, sediment – nil, albumin- +2, sugar and phosphate nil, cast- granular+1, pus cells- wbc 15-20, rbc- 20-25, epithelial cells +1, bacteria -+1. Blood- +2 positive, ketone bodies, bilirubin, nitrites, leucocytes were negative. 24 hours urinary protein was 6642.5 mg/24 hours. Urine culture and sensitivity was sterile after 48 hrs of incubation at 37 degree centigrade. Serum lipid profile- serum cholesterol- 468.2 mg/dl, serum triglycerides- 350mg/dl, HDL-31.9, LDL- 340.3 mg/dl. Hepatitis B, Hepatitis C, HIV 1&2 were negative. USG (W/A) was s/o moderate to gross ascites, bilateral minimal pleural effusion, b/l altered renal echogenicity with normal size and maintained corticomedullary differentiation. 2D-Echo was within normal limits. Renal biopsy was send and was suggestive of mesangial and focal endocapillary proliferative IgA Nephropathy. In view of nephrotic range proteinuria and other features suggestive of nephritic syndrome pt was started on high dose intravenous steroids (1gm methylprednisolone/ day for 5 days).Following steroid therapy the patient improved drastically. The anasarca resolved, renal function improved, and proteinuria decreased. Day 7 24 hours urinary protein reduced to 2.3 gms/24 hours, edema subsided, serum creatinine became 0.8 mg/dl and urea 35 mg/dl. Patient was discharged on predisone 1mg/kg/day, ACE Inhibitor –ramipril 5 mg /day. Patient was reviewed after 15 days. 24 urinary protein decreased to 190 mg/24 hours, there was no edema, renal function tests were within normal limits.

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Table 1: Various tests conducted

Hb(gm/dl)	15.2
TLC	14500
DLC	P88L8M4E0
Platelet count	311000/microlt.
Serum Protein(gm/dl)	1.7
Serum Albumin(gm/dl)	4.6
Alkaline phosphate(U/lt.)	264
Serum creatinine (mg/dl)	1.7
BUN (mg/dl)	119.8
Serum Na/k/Ca	
24 hour urinary protein	6642.5 mg
ANA levels (N-<0.8),	0.27
Anti ds-DNA (N-<35.0 IU/ML),	0.6
C3(N- 79-152 mg/dl), /C4(N-16-38 mg/dl	102/ 30.7
Anti GBM (N-<12 U/ML	
c-ANCA (N-<9.6U/ml)/p-ANCA (n- < 6.7 u/ml)	3.0 / 3.0
ASO Titre (N-<116 iu/ml)	55.7
IgA levels (N-82-453 mg/dl).	254
Serum cholesterol	468.2mg/dl
Serum triglycerides	350mg/dl
Renal biopsy	Mesangial and focal proliferative IgA nephropathy, featuring partial cellular crescents over 2/34 (5.8%) glomeruli and segmental secondary sclerosis in 4/34 (11.7%) capillary tufts. Patchy mild acute tubular injury is observed. *MEST-C scores:* mesangial hypercellularity (M score> 0.5)- M1, endocapillary cellularity present- E1, segmental sclerosis present- S1, tubular atrophy or interstitial fibrosis (<25%)- T0, crescents (cellular/ fibrocellular) (<25%)-C1. IgA- 3+mesangial: granular/confluent, IgG- Negative, IgM- Negative,C3/C1q- Negative, Kappa light chains- 1+mesangial,Lambda light chains- 3+mesangial.

DISCUSSION

IgA nephropathy (IgAN) is the most common primary glomerulonephritis worldwide (D'Amico, 1987; Julian *et al.,* 1988). B-The dominant mesangial IgA deposits represent the diagnostic hallmark of IgAN(3,4). Although nephrotic-range proteinuria is not uncommon in IgAN patients, particularly those with poorly controlled hypertension, a complete nephrotic syndrome is distinctly uncommon. In such cases, coincidental IgAN with minimal-change nephropathy should be excluded by electron microscopy and, if present, treatment should be analogous to minimal-change disease. A rapidly progressive and crescentic IgA nephropathy (IgAN) is uncommon, but it has a high risk of progression to end-stage renal disease and variable response to immunosuppression. The importance of a positive anti-neutrophil cytoplasmic antibody (ANCA) serology in this group of patients is not fully understood but may have prognostic significance. On the other hand, there is growing evidence of the role of complement in the pathogenesis of IgAN, especially in cases of crescentic IgAN (Habib and Jennette, 2006). The rate of IgAN progression is very variable, but after 25 years of follow-up, 30–50% of patients develop end-stage renal disease (ESRD) (Coppo and D'Amico, 2005). The presence of minimal (<500 mg/day) or no proteinuria generally indicates a good long-term prognosis,whereas hypertension is a contributing factor for the progression of renal impairment. Acute kidney injury can accompany gross haematuria episodes; renal function recovery is usually observed after the disappearance of macrohaematuria, but it can be incomplete in elderly patients and in those with prolonged macrohaematuria bouts (>10 days). A particularly rapid progression to ESRD has been reported in a minority of IgAN patients (<10%). Histopathological findings in these patients are in some instances reminiscent of vasculitis, due to the presence of cellular/fibrous crescents, fibrinoid necrosis and arteriolar damage.

Treatment: Although nephrotic-range proteinuria is not uncommon in IgAN patients, particularly those with poorly controlled hypertension, a complete nephrotic syndrome is distinctly uncommon. In such cases, coincidental IgAN with minimal-change nephropathy should be excluded by electron microscopy and, if present, treatment should be analogous to minimal-change disease. Patient was given Regimen of i.v. bolus injections of 1 g methylprednisolone for 3 days followed by oral steroid 0.5 mg/kg prednisone on alternate days. Alternatively the patient could also be given 6-month regime of oral prednisone starting with 0.8–1 mg/kg/d for 2 months and then reduced by 0.2 mg/kg/d per month for the next 4 months.

Conflict of Interest: No potential conflicts of interest
No research involving Human Participants and/or Animals

REFERENCES

Barratt J (2007). Immunopathogenesis of IgAN. *Semin Immunopathology* **29** 427–443.

Coppo R D'Amico G (2005). Factors predicting progression of IgA nephropathy. *Journal Nephrology* **18** 503–512.

D'AmicoG (1987). The commonest glomerulonephritis in the world: IgA nephropathy. *Q Journal of Medicine* **245** 709–727.

Freda BJ, Braden GL (2012). How benign is IgA nephropathy with minimal proteinuria? *Journal of American Society of Nephrology* **23** 1607-10.

Habib R Jennette JC (2006). Immunoglobulin A nephropathy and Henoch-Schonlein purpura. *In:* Fogo AB Bruijn JA Cohen AH Colvin RB Jennette JC (eds). *Fundamentals of Renal Pathology New York*: *Springer*, 61–69.

Julian BA, Waldo FB, Rifai A, Mestecky J (1988). IgA nephropathy, the most common glomerulonephritis worldwide. A neglected disease in the United States? *American Journal of Medicine*; 84 129-32.