Case Report

A Case Study on Berger's Disease

Sonia Singh[1], S Sai Balaji Iyer[2], Prithvika wagle[3]

1 Asst. Professor, Department of Pharmacy Practice, College of Pharmaceutical Sciences, Dayananda Sagar University, Bengaluru -560078, India.
2,3 Interns Pharm.D, Department of Pharmacy Practice, College of Pharmaceutical Sciences, Dayananda Sagar University, Bengaluru -560078, India.
***Corresponding author's E-mail:** soniapb6895@gmail.com

Received: 18-01-2023; **Revised:** 20-02-2023; **Accepted:** 27-02-2023; **Published on:** 15-03-2023.

ABSTRACT

IgA nephropathy, also known as Berger's disease, is characterized by predominant IgA deposition in the glomerular mesangium. A patient of 31-year-old male who is a known case of CKD stage 5 on regular Haemodialysis, IgA nephropathy seizure disorder, and Hypertension (for seven years) on treatment presented to the emergency. The confirmatory diagnosis was an acute febrile illness, lower respiratory tract infection, bilateral pneumonitis, severe anaemia, chronic kidney disease stage 5, IgA nephropathy, seizure disorder, and accelerated hypertension. Meanwhile, he was dialyzed adequately as a life-saving treatment along with other life supportive measures.

Keywords: : IgA, Berger disease, Nephropathy, Haemodialysis, Glomerular mesangium.

QUICK RESPONSE CODE →

DOI: 10.47583/ijpsrr.2023.v79i01.009

DOI link: http://dx.doi.org/10.47583/ijpsrr.2023.v79i01.009

INTRODUCTION

IgA nephropathy, also known as Berger's disease, is characterized by predominant IgA deposition in the glomerular mesangium. In 1968, Berger and Hinglais provided the first description. It is one of the most typical causes of glomerulonephritis worldwide[1]. Most IgA nephropathy cases are idiopathic, but a respiratory tract infection frequently precedes the onset or worsening of the condition. There have been reports of associations with certain bacteria, including Haemophilus influenzae. Other reasons include liver cirrhosis, gluten enteropathy (celiac disease), HIV infection, and familial IgA nephropathy [2]. Patients with IgA nephropathy may not present any symptoms, yet they frequently have high blood pressure, chronic microscopic hematuria, and proteinuria. Adults are the most common demographic for this presentation. In such circumstances, renal function may be impaired, and remission is unusual. Proteinuria (less than 3 gm/day more common), nephrotic syndrome (more than 3.5gms of proteinuria with edema, hypoalbuminemia, hypertension, and hyperlipidemia), and chronic renal failure are among the symptoms of the condition[3].

Starting with galactose-deficient IgA1, which has less than a full complement of galactose residues on the O-glycans in the hinge region of heavy chains, IgA nephropathy appears to be the result of an ordered series of events. These may function as auto-antigens that cause the development of glycan-specific antibodies and the synthesis of circulating immune complexes that are deposited in the renal mesangium. By releasing pro-inflammatory cytokines, secreting chemokines, and causing macrophages to migrate into the kidney, they eventually cause glomerular damage. Immune complexes, including galactose-deficient IgA and IgG, cause glomerular deposition [4]. The only medications now available to alter mesangial IgA deposition are those that have been extrapolated from the therapy of other forms of chronic glomerulonephritis. Since there are still no randomized controlled trials in IgA Nephropathy pertinent to contemporary clinical practise, there is still no consensus on the use of immunosuppressive drugs for treating progressive IgA Nephropathy. No specific treatment is needed for patients with recurrent macroscopic haematuria or isolated microscopic haematuria and proteinuria <1 g/24 h. Patients with nephrotic syndrome and minimal change on a renal biopsy should be treated as those with minimal change nephropathy. The use of corticosteroids for nephrotic IgA Nephropathy outside this subset of individuals is not supported by any data [5].

Case Presentation

A patient of 31-year-old male who is a known case of CKD stage 5 on regular Hemodialysis, IgA nephropathy seizure disorder, and Hypertension (for seven years) on treatment presented to the emergency with complaints of acute febrile illness, breathlessness with cough, easy fatiguability and accelerated hypertension (200/120 mm Hg at the time of admission) requiring ICU admission. On admission, relevant investigations are done. On physical examination, he appeared to be extremely pale. Vitals signs were: HR (144 bpm), BP (200/120 mm Hg), SpO_2: 84% on RA (95% with oxygen support), and Temperature: 103^0F. The patient had a fever, tachycardia, accelerated hypertension,

and shortness of breath. On systemic examination, bilateral basal crepts were found in the respiratory system. The blood report showed severe anaemia with Hb-4.9g/dl. Immediately, three units of PRBC were transfused and put on dialysis. Correction of Vitamin B12(114) was given. Peripheral smear showed early macrocytic anaemia with relative neutrophilia. HRCT results showed extensive patchy areas of ground glass opacifications with air space and opacification in the bilateral lung parenchyma (left >right side), suggesting pneumonitis. 2D ECHO showed tachycardia (+), no definitive RWMA, and suitable LV systolic function with LVEF- 55%. The confirmatory diagnosis was an acute febrile illness, lower respiratory tract infection, bilateral pneumonitis, severe anaemia, chronic kidney disease stage 5, IgA nephropathy, seizure disorder, and accelerated hypertension. He was treated with appropriate IV antibiotics, antacids, nebulization, Oxygenation, and other supportive measures during the hospital stay. Meanwhile, he was dialyzed adequately as a life-saving treatment. As the patient was symptomatically better and hemodynamically stable, he was discharged with advice. The progress chart of the patient is as follows:

Day 1 (28/09/2022): At 1 PM, the patient's vitals were as follows: HR (144 bpm), BP (200/120 mm Hg), SpO_2: 84% on RA (95% with oxygen support) and Temperature: 103^0F and severely anemic, his Hb level being a mere 4.9 gm/dL. Following this, he was admitted to ICU and was given Inj. MAGNEX FORTE 1.5 gms IV BD, Tab. DOLO 650 mg, Neb. DUOLIN Q6H and Neb. BUDECORT Q8H along with 1st dose of inj. Vancomycin 1 gm and Inj. Amikacin 500 mg. Arrangement of 3 units of PRBC transfusion was done. The patient was then advised for USG abdomen, chest X-ray, and 2D ECHO. At 4:30 PM, 2D ECHO reports indicated that the patient was tachycardic with a heart rate of 138 bpm and concentric LVH. Chest X-Ray reports showed bloodstream infection with fluid overload, and B/L quad infiltrates suggestive of pulmonary edema with B/L severe crepts and rhonchi sounds being prominent. At 5 PM, arterial blood gas analysis was done: HCO3 (35.9), Spo2 (96.2%), BF (1.6), pH (7.49), PaO_2 (77.7), $PaCO_2$ (32.2), Lactate (0.4), Sodium (142 mmol/L) and potassium (4.1 mEq/L). The patient was kept on a ventilator with NIV FiO2 40 % (2:2: HS).

Day 2 (29/09/2022): At 8:30 AM, the patient reviewed symptomatically with complaints of cough and worked breathing reduced. Vitals were as follows: HR (108 bpm), BP (190/120 mm Hg), SpO_2 (96% with 2L Oxygen), and afebrile, RS: Crepts were heard along with occasional rhonchi, and he was conscious and oriented. To discuss HRCT films, the patient was advised to RT-PCR/H1N1 swab and repeat ECHO when HR <100 bpm. At 9:10 AM, the vitals were: HR (132 bpm), BP (175/96 mm Hg), and SpO_2 (96% with 5L oxygen). Laboratory investigations were as follows: Hb (7.86), Chloride (111 mmol/L), LDH (382), decreased Vit B12 (114 pg/ml), raised creatinine and reticulocyte levels (4.60 mg/dL and 3.5% respectively). The patient was stool examined for occult blood and administered Inj. ELDERVITA 12 mg OD for three days (weekly once), 1 unit of PRBC during hemodialysis along with leucocyte filter/UF 1000/, Inj. AMIGO PLUS, Tab. DOLO 650 mg SOS. The patient was severely anemic (Hb-7.80), ECG and peripheral smear showed sinus tachycardia and early macrocytic anemia with neutrophilia, vitals were stable, cough and dyspnea reduced, SpO_2 (95% with 2L oxygen), chest X-Ray film showed non-dependent ground glass opacities suggestive of atypical pneumonia. The patient was given Cap. Tamiflu 75 mg OD. At 9:45 PM, Hb (7.8 gm/dL), HR (121 bpm), and RT-PCR reports were negative. CT score was 15/25, and HRCT showed multiple patchy areas of ground glass opacification. USG of abdomen reports showed B/L small kidneys with grade 2 medical renal disease with the right and left kidney measuring 7.8 cms* 70 mm and 8 cm* 80 mm, respectively, mild parenchymal thinning, and moderate increased cortical echotexture.

Day 3 (30/09/2022): At 9:15 AM, the measured vitals were as follows- HR (112 bpm), BP (153/106 mmHg), SpO_2 (97%), afebrile temperature and Hb was 8.69. The patient was planned for proctoclysis enema, hemodialysis with a high protein diet, SLED/4hrs/saline, no heparin/UF 1000, 1 unit of PRBC with leucocyte filter during hemodialysis, and Inj. AMIGO PLUS 200 mg IV during hemodialysis. At 4:30 PM, vitals were as follows- HR (127 bpm), BP (130/90 mm Hg). The patient was also given a TAB. MINIPRESS XL 10 mg BD, TAB. NICARDIA R 20 mg tid, TAB. CARDIVAS 5 mg P/O, SYP. LACTULOSE 15 ml P/O BD and TAB. DOLO 650 mg Q6H SOS.

Day 4 (01/10/2022): At 9:50 AM, the vitals were as follows- HR (105 bpm), BP (140/90 mm Hg), SpO_2 (95% on RA), and Hb level was 8.90. Hemodialysis was done with potassium dialysate 4 mEq, Inj. KCl 20 mEq IV during hemodialysis in 100 ml over 3 hours. Inj. AMIGO PLUS (1-unit PRBC during HD) with Inj. VITNEURIN 1 amp in 50 ml and Inj. VANCOMYCIN 50 ml over 15 hrs, Inj. PAN 40 mg IV OD, TAB. MINIPRESS XL 10 mg BD, TAB. NICARDIA R 20 mg tid, TAB. CARDIVAS 5 mg P/O, SYP. LACTULOSE 15 ml P/O BD and TAB. DOLO 650 mg Q6H SOS, NEB. DUOLIN, NEB. BUDECORT, TAB. LEVIPIL 50 mg P/O BD, TAB. SELBO 400 mg 400 mg BD, Inj. MEROPENEM 500 mg IV, Inj. LEVODAY 250 mg IV (in 50 ml dilution) OD.

Day 5 (02/10/2022): At 10:30 AM, the vitals were as follows- HR (126 bpm), BP (160/100 mm Hg), SpO_2 (97%), and Hb level was 9.22. The patient was administered Inj. VITNEURIN IV in 50 ml NS over 1 hour. He was given Inj. PAN 40 mg IV OD, TAB. MINIPRESS XL 10 mg BD, TAB. NICARDIA R 20 mg tid, TAB. CARDIVAS 5 mg P/O, SYP. LACTULOSE 15 ml P/O BD and TAB. DOLO 650 mg Q6H SOS, NEB. DUOLIN, NEB. BUDECORT, TAB. LEVIPIL 50 mg P/O BD, TAB. SELBO 400 mg 400 mg BD, Inj. MEROPENEM 500 mg IV, Inj. LEVODAY 250 mg IV (in 50 ml dilution) OD.

Day 6 (03/10/2022): At 9:30 AM, the vitals were as follows- HR (80 bpm), BP (170/110 mm Hg), Hb (8.83 gm/dL), raised S. Creatinine (6.8 mg/dL) and raised chloride levels (112 mmol/L). He was given Inj. PAN 40 mg IV OD, TAB. MINIPRESS XL 10 mg BD, TAB. NICARDIA R 20 mg tid, TAB. CARDIVAS 5 mg P/O, SYP. LACTULOSE 15 ml P/O BD and

TAB. DOLO 650 mg Q6H SOS, NEB. DUOLIN, NEB. BUDECORT, TAB. LEVIPIL 50 mg P/O BD, TAB. SELBO 400 mg 400 mg BD, Inj. MEROPENEM 500 mg IV, Inj. LEVODAY 250 mg IV (in 50 ml dilution) OD. The patient was planned for discharge.

DISCUSSION

The most prevalent cause of end-stage renal disease in people with primary glomerulopathy is IgA nephropathy, the most prevalent form of primary glomerulonephritis worldwide [6]. During the second and third decades of life, the incidence significantly rises [7]. It is recognized that deposition of IgA with a majority of Lambda light chains occurs in the renal mesangium, though the pathogenesis is not entirely understood[8]. Patients typically present with gross haematuria following an upper respiratory tract infection or they may present with microscopic haematuria with varying degrees of proteinuria[6]. In more severe cases, nephrotic-range proteinuria is frequently observed. Following an episode of a non-specific upper respiratory tract infection, episodic haematuria is the characteristic sign[10]. IgA nephropathy exhibits a wide range of clinical and pathological features. Clinical features range from asymptomatic haematuria to Rapidly Progressive Glomerulonephritis (RPGN). Clinically, RPGN is indicated by a sudden drop in glomerular filtration rate (GFR) of at least 50% over a brief time, ranging from a few days to three months [11]. Thrombotic microangiopathy can also be found in IgA nephropathy due to malignant hypertension and indicates a poor renal outcome, as seen in our case. Increased serum creatinine (Cr), decreased glomerular filtration rate (GFR), hypertension (blood pressure > 140/90 mmHg), and chronic proteinuria are some of the clinical and laboratory findings that might assist in stratifying the severity of the disease at the time of diagnosis [12].

Initial and subsequent renal function may be within normal ranges for a significant time [11]. Renal biopsy is the gold standard for diagnosis since it reveals mesangium proliferation with IgA deposits under immunofluorescence and electron microscopy[13]. The development of ESRD (end-stage renal disease), which affects 15% to 20% of patients by the age of 10, and 20% to 50% of patients by the age of 20, has been shown in long-term follow-up studies of the biopsy-verified cases[14]. Patients with greater systolic blood pressure, lower GFR, haemoglobin, and serum albumin levels at baseline are more at risk of developing ESRD, according to a study by Xie J et al. [15].

CONCLUSION

IgA Nephropathy is a common glomerular disease and a significant cause of renal failure. Furthermore, it may rarely initially just exhibit ESRD-related hypertensive emergency symptoms. Patients are likely to experience a progressive course that results in ESRD; hence early detection and intervention are crucial.

REFERENCES

1. Fabiano RC, Pinheiro SV, Simões e Silva AC. Immunoglobulin A nephropathy: a pathophysiology view. Inflammation Research. 2016 Oct;65(10):757-70.
2. Rollino C, Vischini G, Coppo R. IgA nephropathy and infections. Journal of nephrology. 2016 Aug;29(4):463-8..
3. Pattrapornpisut P, Avila-Casado C, Reich HN. IgA nephropathy: core curriculum 2021. American Journal of Kidney Diseases. 2021 Sep 1;78(3):429-41.
4. Knoppova B, Reily C, Maillard N, Rizk DV, Moldoveanu Z, Mestecky J, Raska M, Renfrow MB, Julian BA, Novak J. The origin and activities of IgA1-containing immune complexes in IgA nephropathy. Frontiers in immunology. 2016 Apr 12;7:117.
5. Barratt J, Feehally J. Treatment of IgA nephropathy. Kidney international. 2006 Jun 1;69(11):1934-8.
6. Donadio JV, Grande JP. IgA nephropathy. New England Journal of Medicine. 2002 Sep 5;347(10):738-48.
7. Simon P, Ramee MP, Boulahrouz R, Stanescu C, Charasse C, Seng aNG KI, Leonetti F, Cam G, Laruelle E, Autuly V, Rioux N. Epidemiologic data of primary glomerular diseases in western France. Kidney international. 2004 Sep 1;66(3):905-8.
8. Roberts IS. Pathology of IgA nephropathy. Nature Reviews Nephrology. 2014 Aug;10(8):445-54.
9. Donadio JV, Grande JP. IgA nephropathy. New England Journal of Medicine. 2002 Sep 5;347(10):738-48
10. Chang A, Kowalewska J, Smith KD, Nicosia RF, Alpers CE. A clinicopathologic study of thrombotic microangiopathy in the setting of IgA nephropathy. Clinical nephrology. 2006 Dec 1;66(6):397-404.
11. Wyatt RJ, Julian BA. IgA nephropathy. New England Journal of Medicine. 2013 Jun 20;368(25):2402-14.
12. Lv J, Yang Y, Zhang H, Chen W, Pan X, Guo Z, Wang C, Li S, Zhang J, Zhang J, Liu L. Prediction of outcomes in crescentic IgA nephropathy in a multicenter cohort study. Journal of the American Society of Nephrology. 2013 Dec 1;24(12):2118-25.
13. Geddes CC, Rauta V, Gronhagen-Riska C, Bartosik LP, Jardine AG, Ibels LS, Pei Y, Cattran DC. A tricontinental view of IgA nephropathy. Nephrology Dialysis Transplantation. 2003 Aug 1;18(8):1541-8.
14. Walsh M, Sar A, Lee D, Yilmaz S, Benediktsson H, Manns B, Hemmelgarn B. Histopathologic features aid in predicting risk for progression of IgA nephropathy. Clinical Journal of the American Society of Nephrology. 2010 Mar 1;5(3):425-30.
15. Xie J, Kiryluk K, Wang W, Wang Z, Guo S, Shen P, Ren H, Pan X, Chen X, Zhang W, Li X. Predicting progression of IgA nephropathy: new clinical progression risk score. PloS one. 2012 Jun 14;7(6):e38904.

Source of Support: The author(s) received no financial support for the research, authorship, and/or publication of this article.

Conflict of Interest: The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.