Bangladesh Med J. 2022 Sept; 51(3)
DOI: https://doi.org/10.3329/bmj.v51i3.70140

Case Report

IgA Vasculitis in an Adult Man- a Case Report

***Mahin T[1], Faria L[2], Mottaleb AKMA[3], Shumy F[4], Qader MA[5]**

Abstract:

IgA vasculitis is rare in adult. A significant portion of adult cases may present with renal involvement. Although most of these cases can be managed symptomatically, some cases may require immunosuppressive treatment. Unless diagnosis and appropriate treatment is initiated early IgA nephropathy in adult may result in chronic kidney disease. Immunoglobulin A vasculitis (IgAV) is an acute disorder causing generalized vasculitis principally involving skin, gastrointestinal (GI) tract, kidneys and joints. Here, a 28 year old man was presented with purpuric rash with leg ulcer, inflammatory arthritis involving multiple joints, abdominal pain and vomiting. He had mild pedal edema and normal blood pressure. Investigations showed raised inflammatory markers, haematuria, and nephrotic range proteinuria with normal liver and renal function. Biochemical and immunological evaluation of lupus nephritis, ANCA associated vasculitis and other causes of glomerulonephritis were negative. Renal biopsy with direct immunofluorescence showed features of IgA nephropathy. Patient was treated with steroid and mycophenolate mofetil with a good response.

Keywords: *IgA vasculitis, leg ulcer, prednisolone, mycophenolate mofetil.*

1 *Dr. Tasnova Mahin, Associate Consultant, Department of Internal Medicine, Square Hospitals Limited, Panthapath, Dhaka, Bangladesh. Email:drtasnova@squarehopsital.com
2 Dr. Labeeba Faria, Specialist, Department of Nephrology, Square Hospitals Limited, Panthapath, Dhaka, Bangladesh.
3 Dr. AKM Abu Mottaleb, Associate Consultant, Department of Nephrology, Square Hospitals Limited, Panthapath, Dhaka, Bangladesh.
4 Dr. Farzana Shumy, Associate Consultant, Rheumatology, Square Hospitals Limited, Panthapath, Dhaka, Bangladesh.
5 Dr. Md Abdul Qader, Associate Consultant, Paediatric Nephrology, Square Hospitals Limited, Panthapath, Dhaka, Bangladesh.
*For correcpondence

INTRODUCTION

IgA vasculitis is the most common vasculitis in childhood with incidence decreasing with age.[1] The occurrence of IgAV in adults is approximately 3.4- 14.3 cases per million.[2,3] It commonly involves skin, gastrointestinal tract, kidneys, joints and rarely lungs and central nervous system (CNS).[1,4] There is slightly male predominance with a male to female ratio of 1.5-2:1, but some studies have found equal distribution in both sexes.[5,6] It is typically an acute self-limiting illness usually managed with supportive treatments. However approximately 7% of all cases IgAV may present with nephritis or nephrotic syndrome which may progress to chronic kidney disease.[7] What makes this case unique is here we present a case of IgA vasculitis with nephropathy in an adult who needed management with immunosuppressive therapy and had a good response.

CASE HISTORY:

A 28-year-old non-diabetic normotensive gentleman presented with multiple purpuric rash along with leg ulcer for 20 days which was initially reddish painful nodules in the lower limb and subsequently became ulcerative. He also had pain and swelling of right knee, both ankle and small joints of right hand for same duration. He also complained of abdominal pain and vomiting for 2-3 days. He denied any history of fever, bloody diarrhea, and flu like symptoms or urinary symptoms in last 3 months. He had no history of taking any antibiotics or any nephrotoxic drugs prior to this illness. On examination, he had mild ankle edema and his blood pressure was normal. Along with ulcerative lesion in both lower limbs he had petechial rash behind legs, buttock and upper limbs (Figure 1). His initial lab reports revealed mild normocytic normochromic anemia with raised inflammatory markers, renal functions and liver functions were normal. Urine microscopy showed plenty of RBC and a 24 hour urinary total protein was 7.39gm/day (Table 1).

Table- I: Initial lab results of the patient

Characteristics	Results
CBC	Haemoglobin-11.9 g/dl, Leukocyte-14,200/mm^3,Platelet-389,000/mm^3
ESR	37 mm in 1st hour
C- reactive protein	45.6 mg/L
Serum creatinine	0.9 mg/dl
ALT	49U/L
PT and APTT	normal
Urine R/E	P.C-30-35/HPF, RBC-Plenty/HPF, protein- 3+
24 hour urinary total protein	7.39 gram/24 hours
USG of abdomen	Fatty liver (grade-2)

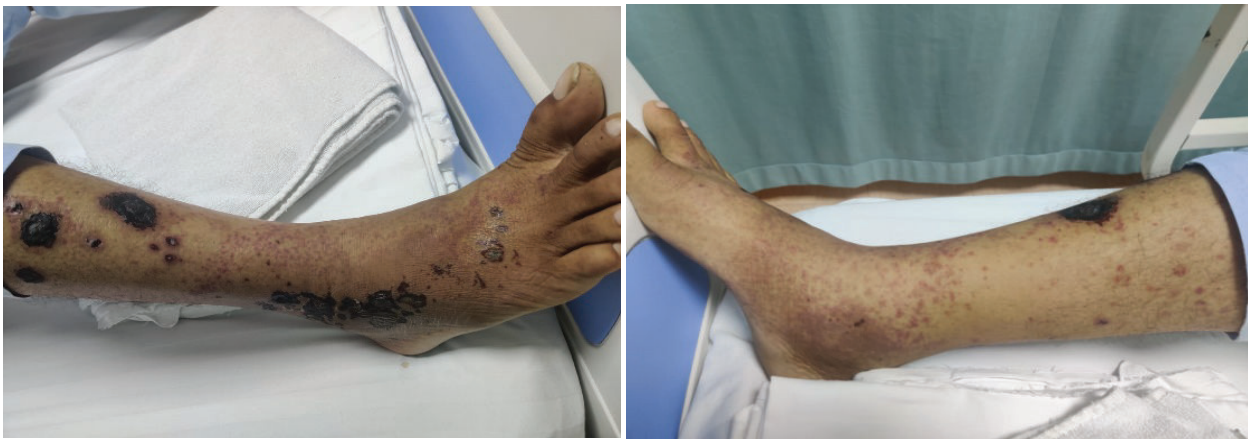

Figure- 1: *Purpuric rash in the lower limb including ulceration*

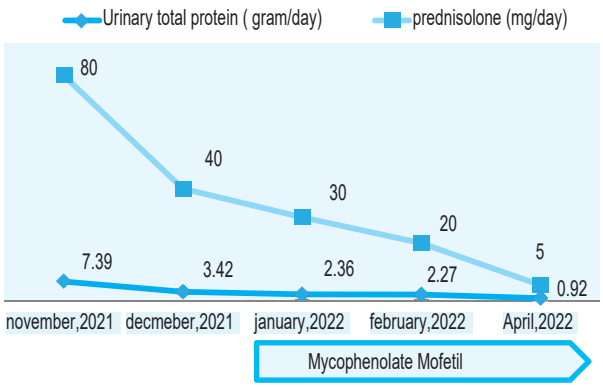

Figure-2: *Line chart showing response of proteinuria to treatment*

His serum complements including C3, C4 level were within normal limit, antinuclear antibody, RA factor, Anti-CCP antibody, C-ANCA, P-ANCA, hepatitis B and C serology were negative. Patient was diagnosed as a case of IgA vasculitis with Nephritis and a renal biopsy was done which revealed mild to moderate global mesangial cell hyperplasia and deposition of IgA, C3 and C1q in immunoflurescence microscopy. He was started on oral prednisolone with gradual tapering along with oral Losartan potassium. Mycophenolate mofetil was added for persisting proteinuria after 2 months. Within 5 months his skin ulcers disappeared and proteinuria reduced to 0.92 gm/24 hours (Figure 2).

DISCUSSION

IgA vasculitis is usually more severe in adults with tetrad of non-thrombocytopenic palpable purpura, arthritis/ arthralgia, gastrointestinal and renal involvement. Cutaneous manifestation may also include urticarial and macular lesion, rarely, hemorrhagic bullae and ulcerative lesion[8]. In our case patient had palpable purpura and skin ulcer. Arthritis and arthralgia may involve 74% of the patients, joints may be swollen, tender and painful but usually no erythema or effusion. Knees and ankles are most

commonly affected; rarely fingers and wrist may be involved[9]. Our patient had involvement of knee joint, ankle joint and small joints of right hand. IgA nephropathy in adult is frequent, affecting from 45%t to 85% of patients.[10] The risk of progression to renal insufficiency is 5-30%.[8,11] Both hematuria and nephrotic range proteinuria as in our case may be present in 29% cases.[12]

Renal Biopsy may reveal mesangial proliferation, leucocyte migration, crescent formation along with deposition of IgA and C3.[13] In our patient, the pathological change in the light microscope was similar and there was IgA and C3 deposition on immunoflurescence.

IgAV is self-limiting in nature in up to 89% adults which can be managed with symptomatic treatment [14]. Oral steroid is recommended in patients with severe rash, renal involvement, severe colicky abdominal pain or scrotal /testicular involvement[8]. In a few studies steroids was found to induce complete remission of nephritis.[15] Floege and feehally adapting KDIGO guideline suggest 6 months therapy of steroid if proteinuria >1 gram/day persist despite renin-angiotensin receptor blockage and blood pressure control.[16] Bernardio et al, reported a case of IgAV in an adult patient with SLE mild proteinuria who was treated with colchicines, steroid and azathioprine with good outcome.[17]

Hocevar et al did an analysis of baseline clinical features and co-morbidities of adult IgAV cohort over 9 year period. It showed hypertension, diabetes and obesity were more prevalent in patients with IgAV then age matched controls. In the study population kidney, GI and articular involvement due to IgAV was 45%, 30.2% and 38.5% respectively. 10% patient was diagnosed with acute kidney injury and arterial hypertension concurrently with IgAV.[18]

According to Yaseen et al. clinical course of IgA vasculitis in adult is different from paediatric population due to its high risk of evolving to end stage renal disease. There is still controversy regarding immunosuppressive therapy in severe IgAV. Recent studies are providing data regarding potential benefits of targeting mucosal immune system, toll- like receptors, complements and tyrosine kinase inhibitors in management of IgA nephropathy. Still there is paucity of high quality evidence based guidelines in management of severe IgAV.[19] .

CONCLUSIONS

IgA vasculitis is rare in adult. Around 5-30% patient with IgA vasculitis with renal involvement may progress to chronic kidney disease. Early diagnosis and treatment is of prime importance in this case.

ACKNOWLEDGEMENT:

Dr. Tasnova Mahin and Dr. Md. Abdul Qader prepared the manuscript. Dr. AKM Abu Mottaleb, Dr. Labeeba Faria and Dr. Farzana Shumy gave their valuable intellectual contribution during diagnosis, management and follow up of the patient and valuable input during preparation of manuscript.

REFERENCES

1. Leung AKC, Barankin B, Leong KF. Henoch-Schonlen Purpura in children: An Updated Review. CurrPediatr Rev.2020;16(4):265-276
2. Saulsbury F. Epidemiology of Henoch-Schonlein purpura. Cleve Clin J Med. 2002;69 (suppl 2):87-9
3. Watts RA, Scott D. Epidemiology of the vasculitides. Semin Resp crit care. 2004;25:455-64
4. Blanco R, Martinez-Taboada VM, Rodriguez-Valverde V, Garcia Fuentez M, Gonzalez-gay MA. Henoch-Schonlein purpura in childhood: epidemiological and clinical analysis of 150 cases over 5-year period and review of literature. Semin Arthritis Rheum.2005 dec; 35 (3):143-53.
5. Gonzalez LM, Janniger CK, Schwartz RA. Pediatric Henoch-Schonlein purpura. Int J Dermatol.2009 nov; 48 (11):1157-65.
6. Lei WT, Tsai PL, Chu SH, Kao YH, Lin CY, Fang LC,et al. Incidence and risk factors for recurrent Henoch-schonlein purpura in children from a 16-year nationwide data base. PediatrRheumatol Online J. 2018 apr; 16(1):25.
7. Nielsen HE. Epidemiology of Schonlein-Henoch purpura. Acta Pediatr Scand.1998;77: 125-131.
8. Fogazzi GB, PasQuali S, Moriggi M et al. Long term outcome of Schonlein-Henoch nephritis in the adult. Clin nephrol1989; 31:60-6.
9. Trapani S, Michei A, Grisolia F, Resti M, Chiappini E, Falcini F et al. Henoch Schonlein purpura in childhood: epidemiological and clinical analysis of 150 cases over 5-year period and review of literature. Semin Arthritis rheum.2005 Dec; 35 (3):143-53.
10. Rieu P, Noel LH. Henoch-Schonlein nephritis in children and adults, Morphological features and clinicopathological correlations. Ann Med Interne 1999; 150:151-9.

11. Goldstein AR, White RH, Akuse R, Chantler C. Long term follow up of childhood Henoch-Schonlein nephritis. Lancet.1992 feb; 339(8788):280-2.

12. Hillhorst M, Paasen P, Van Breda P, Cohen Tervaert JW. Immune complexes in acute adult onset Henoch-Schonlein nephritis. Nephrol Dial Transplant.2011; 26(12); 3960-7.

13. Roberts PF, Waller TA, Brinker TM, Riffe IZ, Sayre JW, Bratton RL. Henoch-Schonlein purpura: a review article. South Med J.2007 Aug; 100(8):821-4.

14. Sohagia A, Gunturu S, Tong TR, Hertan HI. Henoch-schonlein purpura- a case report and review of literature. Gastroenterol Res Pract.2010; 2010:597648.

15. Floege J, Feenhally J. Treatment of IgA nephropathy and Henoch-Schonlein nephritis. J Nat Rev Nephrol. 2013; 9:320-7.

16. Sergura T, Borrego U, Perez Del Barrio P, Ruiz Avila I. Complete remission of nephrotic syndrome with methylprednisolone pulses in an adult with Schonlein- Henoch purpura. Nefrologia.2007; 27(1):96-8.

17. Bernardino V, Mendes-Bastos P, Rodrigues A, Riso N. IgA Vasculitis (formerly Henoch Schonelin purpura) in an adult with systemic lupus erythematosus. BMJ Case Rep.2015: bcr 2015210121.

18. . Hocevar A, Tomsic M, Rotal Z. IgA vasculitis in adults: few certainties and many uncertainities. Ann Rheum Dis 2020: 79:e47.)

19. Yaseen K, Herlitz LC, Villa-Forte A. IgA Vasculitis in Adults: A Rare yet Challenging Disease. Currr Rheumatol Rep.2021 Jul 1;23(7):50) input during preparation of manuscript.