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Henoch-Schoenlein Purpura with IgA Nephropathy Treated with Rituximab: A Case Report

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Abstract

Henoch-Schoenlein purpura (HSP) is a distinct subtype of hypersensitivity vasculitis with IgA nephropathy as major complication. We report a case of 8-year old male who presented with recurrent HSP and IgA nephtopathy successfully treated with rituximab leading to complete and sustained remission in skin lesions and renal function.

Keywords: Henoch-Schoenlein purpura; IgA nephropathy; Rituximab.

Introduction

Henoch-Schoenlein purpura (HSP) is an IgA mediated systemic small vessel vasculitis with IgA deposition in vessel walls leading to symptoms involving skin, joints and kidneys. It typically affects children following a respiratory tract infection.[1] HSP is more frequent in two to eight years of age with male predominance (M:F=1.5:1).[2] Although HSP is generally a self-limiting disease, the long-term prognosis depends on the severity of renal involvement. Small subsets of patients are unable to wean off corticosteroids and novel corticosteroid sparing and safe immuno modulators are needed.

Case Report

An eight-year old male child presented with history of recurrent skin lesions on legs associated with fever and joint pains for 4 months (Fig. 1). There was no history of drug intake prior to first episode. Patient was treated with antibiotics and NSAIDs during each episode with recurrence within 15–20 days each time. Patient presented to us during 4th episode with bilateral symmetrical, tender, purpuric lesions on lower extremities associated with edema feet and scrotum.

Baseline laboratory investigations revealed total white blood cell (WBC) count 17,400/cumm with normal differential count. Urine routine microscopy analysis showed; Red blood cells (RBC) 28 cells/ hpf and albumin ++. 24–hour urinary protein was 330mg (N <140mg/day). Platelet count, renal and liver function tests were within normal range. Stool examination for occult blood was negative. The anti-Streptolysin O titter, C-reactive protein, and complement levels were in the normal range. S.ANA, DsDNA and ANCA were negative. HIV, HSV, HbSAg, HCV serology were non-reactive. X-ray chest and abdominal sonography did not

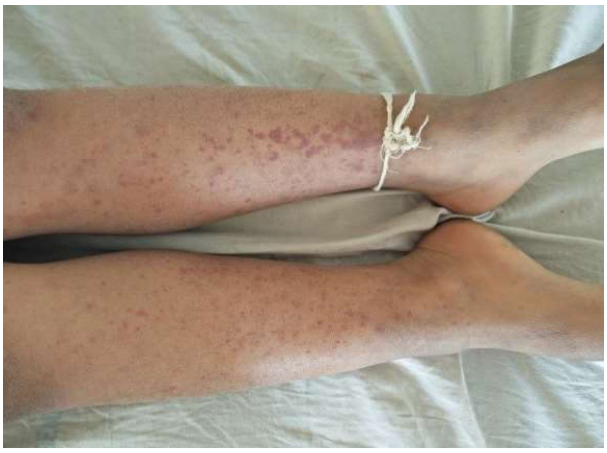

Fig. 1: Palpable Purpura with Oedema of Distal Extremity.

show any abnormality. Skin biopsy on routine histopathology showed an inflammation of small vessels in the upper dermis with a dense infiltrate of neutrophils and fibrin deposits into vascular wall. Direct immuno fluorescence did not show any deposits. USG guided renal biopsy showed localized mesangial proliferation in glomeruli with interstitial mononuclear infiltrate confirming mesangial proliferative glomerulonephritis. Immuno fluorescence showed strong positive IgA and weak positive C3 deposits in the mesangium. Based on clinical, histopathological and renal biopsy findings diagnosis of Henoch-Schoenlein purpura associated with IgA nephropathy (mesangial proliferative glomerulonephritis) was concluded.

Patient was prescribed oral prednisolone 2mg/kg which was tapered to 1.5mg/kg over 20 days. Repeat urine examination at day 20 showed persistent hematuria with RBC 70 cells/ hpf. Clinically fever and edema feet persisted. Nephrologist advised to continue high dose steroid for at least 6 weeks without tapering due to renal hypertension but; as patient was developing side effects of systemic steroids in terms of weight gain; alternative treatment options were searched for and as per references in literature of use of rituximab in pediatric HSP with renal involvement[3,4] and consent from parents, intravenous infusions of Rituximab 400mg (375mg/m2 approximately) in 200ml normal saline twice at 2 weeks interval were given along with tapering dose of corticosteroids. After first rituximab infusion itself patient improved symptomatically with normal temperature and reduction in joint pain. RBC count in urine reduced significantly by day seven (55 cells/hpf). After second infusion rituximab urine examination returned to normal range, total WBC count reduced to 10,400/cumm; scrotal edema, leg edema as well as fever subsided completely. 24-hour urinary protein also returned to normal level. As there was improvement in all parameters, tapering of steroid was started 10 days after 1stinfusion of rituximab and the same was rapidly tapered to 10 mg/day by week two post 2nd infusion. Within 3 weeks of last rituximab infusion; patient was able to stop steroid completely with complete recovery of renal function and no recurrence in one year follow-up.

Discussion

HSP is a systemic vasculitis characterised by the clinical tetrad of palpable purpura, arthritis, haematuria and abdominal pain. The most serious complication is renal involvement which requires treatment with long term corticosteroids. The pathogenesis of the nephropathy involves the deposition of aberrant glycosylated IgA1 and/ or of IgA1 immune complexes in the glomerular mesangium.[5] A meta-analysis has shown that there is no evidence of benefit of prednisone in preventing serious long-term kidney disease in HSP.[6] Thorne et al report three paediatric patients successfully treated with rituximab for severe refractory HSP.[3] Crayne et al also reported steroid sparing effect of rituximab in chronic steroid dependent and immuno modulator refractory HSP in eight paediatric cases.[4] The probable mechanism of action of rituximab in IgA nephropathy is loss of IgA producing B-cells and effect on cytokine production and antigen presentation.[3, 4]

We report here a case of pediatric patient with refractory HSP successfully treated by two rituximab infusions without any adverse events. Our patient had a rapid improvement in skin and renal symptoms after rituximab leading to a dose reduction of corticosteroids without relapse.

Candidates for rituximab treatment could be patients with poor risk factors with progression to end stage renal disease or patients with severe HSP experiencing relapse, as a steroid sparing agent. We recommend more studies of rituximab in patients of HSP with renal involvement.

References

1. Bagai A, Albert S, Shenoi SD. Evaluation and therapeutic outcome of palpable purpura. Indian J Dermatol VenereolLeprol 2001;67:320–3.
2. Bluman J, Goldman RD. Henoch-Schönlein purpura in children: limited benefit of corticosteroids. Can Fam Physician 2014;60(11):1007–1010.

3. Donnithorne KJ, Atkinson TP, Hinze CH, Nogueira JB, Saeed SA, Askenazi DJ, Beukelman T, Cron RQ. Rituximab therapy for severe refractory chronic Henoch-Schönlein purpura. J Pediatr2009;155(1):136–9.
4. Crayne CB, Eloseily E, Mannion ML, Azerf SP, Weiser P, Beukelman T et al. Rituximab treatment for chronic steroid-dependent Henoch-Schonlein purpura: 8 cases and a review of the literature. Pediatric Rheumatology 2018;16:71.
5. Lau KK, Suzuki H, Novak J, Wyatt RJ. Pathogenesis of Henoch-Schonlein purpura nephritis. Paediatric Nephrol 2010;25:19–26.
6. Chartapisak W, Opastirakul S, Hodson EM, Willis NS, Craig JC. Interventions for preventing and treating kidney disease in HSP, Cochrane Database Syst Rev, 2009;8:(3).