**Henoch Schönlein Purpura Mimicking Lupus Nephritis-A Rare Case**

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**Abstract**

Henoch Schönlein Purpura, is the most common non granulomatous, immune complex mediated small vessel vasculitis in children involving multiple organs. HSP nephritis is reported to occur in 20-80% of patients and characterized by mesangial or mesangioproliferative glomerulonephritis with varying degree of hypercellularity. On Immunofluorescence it is characterized by granular deposits of IgA and to lesser extent IgG or IgM. Full house immunofluorescence with deposits of IgG, IgM, IgA, C3, C4, C1q, kappa and lambda is commonly reported in Lupus and is not a characteristic picture of HSP nephritis. Our patient is a seven year old girl who presented with edema over dorsum of hands and feet, arthritis and immune complex mediated rapidly progressive nephritis with full house picture on immunofluorescence.

**Key words**: HSP, lupus, nephritis

**Introduction**

Henoch Schönlein Purpura (HSP), the most common non granulomatous, immune complex mediated small vessel vasculitis in children involves multiple organs [1–2].

In adults and infants less than 2 years it tends to have an atypical course with higher rate of gastrointestinal and renal complications [1]. HSP nephritis is reported to occur in 20-80% of patients and characterized by mesangial or mesangioproliferative glomerulonephritis with varying degree of hypercellularity. Rapidly progressive glomerulonephritis is rare in children with HSP. One to seven percent patients suffer from end stage renal disease [3,4]. On Immunofluorescence renal biopsy shows, predominantly granular deposits of IgA in the mesangium and to a lesser extent IgG or IgM deposits [5]. Full house on immunofluorescence is the term given to the characteristic histological findings in lupus nephritis which include glomerular deposits of IgG, IgM, IgA, C3, C4 and C1q [6]. We report a case of HSP who presented with edema over dorsum of hands and feet, arthritis and immune complex mediated rapidly progressive nephritis with full house picture on immunofluorescence-a rare occurrence.
Case Report

A seven year old female child presented with swelling and pain in bilateral ankles and wrists for 15 days and low grade fever for two days. There was no history of rash, photosensitivity, abdominal pain, hematuria, diarrhea, oliguria or swelling of face & feet. She was hypertensive (BP-148/98mmHg), had pallor and significant cervical lymphadenopathy. Abdomen examination revealed hepatomegaly (liver span 11cm), but no splenomegaly or free fluid. Musculoskeletal examination revealed swelling with an erythematous hue on dorsum of both hand and feet, and grade two swelling and tenderness in both ankles and right wrist. Other systemic examinations were normal. Five days after admission she developed palpable non blanchable purpuric rash on both legs.

Laboratory tests revealed a total leucocyte count of 12900/cumm (P-50, L36), Hb-7.2 gm/dl, platelet count 3.2 lakhs/cumm, ESR-50mm 1st hour, CRP 64mg/dl, urea 42mg/dl and creatinine of 0.6mg/dl. Liver functions, total proteins and cholesterol were normal. Urine examination revealed microscopic hematuria and albuminuria. Twenty four hour urinary protein was 58mg/m2/day. Serum complement-3 was low (925mg/l, normal-1032-1495) while Complement-4 was normal (222mg/l, normal167-385). Anti Nuclear Antibody, Anti ds DNA, Cytoplasmic Anti neutrophilic cytoplasmic antibody and Perinuclear Anti neutrophilic cytoplasmic antibody were negative. Serum IgA levels were raised-362mg/dl (30-240mg/dl). HIV serology and HBsAg were negative. Mono spot test was negative. Renal Doppler Ultrasonography was normal.

Skin biopsy from the rash, showed mild hyperkeratosis and acanthosis with dermis showing karryorrhetic debris and perivascular infiltrates with neutrophils, lymphocytes and eosinophils and fibrin deposit within vessel wall and subcutaneous tissue. Immunofluorescence was suggestive of IgA deposits. The picture was suggestive of leucocytoclastic vasculitis.

Renal biopsy revealed 15 glomeruli, of which 8 showed global and one showed segmental sclerosis. Protein reabsorption granules were seen in tubules with necrotic debris in few. Diffuse lymphoplasmacytic infiltrate were seen in the interstitium (Figure 1A). Eight glomeruli showed complete fibro cellular crescents (Figure 1B,1C). The remaining seven glomeruli showed increase in mesangial matrix and cellularity with focal endothelial cell proliferation (Figure 1D). Basement membrane revealed wrinkling on silver methenamine stain. One glomerulous showed an active area of fibrinoid necrosis of the glomerular tuft (Figure 1E). Blood vessels revealed medial sclerosis. Immunofloveuscence studies showed a full house staining pattern with glomerular deposits of IgG, IgM, IgA, C3, C4, C1q, and fibrinogen.
IgM, IgA, C1q, C3, C4 Kappa and Lambda of almost equal intensity in the mesangium and peripheral capillary walls. Crescents were identified in the fibrogen stain (Figure 1G). The renal biopsy picture was suggestive of focal necrotizing and crescentic glomerulonephritis (immune complex mediated). Four glomeruli showed healed areas of mesangiolysis in the form of segmental sclerosis (Figure 1F).

In view of the clinical and serological findings, skin biopsy and kidney biopsy, the patient was diagnosed as Henoch-Schonlein purpura with crescentic nephritis.

She was given Amlodipine and pulse methyprednisolone 30mg/kg IV (max 1gm) daily for 3 doses followed by oral prednisolone 2mg/kg/day for 4 weeks and tapered to 0.5mg/kg/day and cyclophosphamide 500-750 mg/m² IV every 3-4 weeks for 6 pulses. The child is doing well on follow up and her blood pressure is under control. Urine is free of albumin and red cells and complement 3 levels have normalized.

**Discussion**

Henoch schonlein purpura is characterized by a classical tetrad of non thrombocytopenic palpable purpura, arthritis or arthralgia, gastrointestinal and renal involvement and rarely other systems involvement [7]. Palpable purpura the hallmark cutaneous lesion is seen in almost 50% of the cases as a presenting sign [7,8].

Edema of dorsum of hands and feet has been characteristically reported in infants [9,10]. Joint involvement, mostly large joints have been reported to precede appearance of rash in less than quarter of patients [10].

Renal involvement usually occurs after 3 to 4 weeks of illness though rarely may precede the appearance of purpura [3,4].

Of the spectrum of the renal involvement, rapidly progressive glomerulonephritis is the rarest [4].

Renal pathology in patients with HSP has been characterized by hypercellularity in mesangium and occasional crescents in cases of rapidly progressive glomerulonephritis.

On Immunofluorescence it is characterized by granular deposits of IgA and to a lesser extent IgG or IgM [5]. Our patient showed a full house immunofluorescence picture with deposits of IgG, IgM, IgA, C1q, C3, C4, kappa and lambda, not a characteristic feature of HSP nephritis. Full house picture has been commonly reported in Lupus and in fact even considered a hallmark of it. Hyperactivity of B cells and decreased activity of T suppressor cells, genetic predisposition to Fc receptor and erythrocyte C3B receptor deficiency are thought to play a pathogenetic role in lupus nephritis by promoting deposition of profound circulating immune complexes and defective immune clearance [11]. Few patients have been reported in literature which had no clinical or serological evidence of systemic lupus erythromatosus but had renal limited lupus [12], HIV nephropathy and glomerulonephritis associated with chronic liver disease have also been associated with full house picture in immunofluorescence [13]. Our patient did not have any clinical or serological findings of chronic liver disease or HIV. Although she had features suggestive of arthritis and low C3, she did not meet the four required criteria for SLE. Moreover her ANA and Anti-dsDNA were negative. In addition she developed classical rash of HSP thereby excluding the diagnosis of SLE. Her characteristic clinical features of HSP were supported by skin biopsy but renal biopsy picture resembled that of lupus nephritis. The exact mechanism of full house on immunofluorescence in a child with HSP could not be ascertained.

Our patient had low complement 3 which improved on treatment as reported by other authors [14] however we could not find any correlation of hypocomplementemia with severity of disease or prognosis [8].

We could come across two case reports in literature similar to our case. Al attarch et al reported a 12 year old girl who presented with HSP with severe IgA nephropathy and responded to treatment but eventually developed lupus. The histopathology did not reveal a full house in the child on immunofluorescence [15]. Abdawani et al reported a 13 year old boy with HSP with nephritic range proteinuria and a full house picture on immunofluorescence which improved with aggressive treatment like our patient [16].

Hence our patient was interesting from two points of view, that edema of hands and feet occurring at the age of seven and a full house immunofluorescence pattern in renal biopsy and features of crescentic glomerulonephritis. Association between occurrence of edema over hands and feet in an
older child with HSP and crescentic glomerulonephritis; if any, needs to be explored.

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**References**