CASE STUDY

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# Immunoglobulin - A Vasculitis: A case study

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**ABSTRACT:**

Henoch-schonlein-purpura (HSP) otherwise called immunoglobulin-A vasculitis is the most common form of small vessel vasculitis. It affects the vasculature of several systems including joints, kidneys, gastro-intestinal tract and skin which results in multi-organ dysfunctional involvement. A 12 year old female child was admitted to the paediatric department of the Virudhunagar government medical college hospital with the complaints of red raised stain lesions over the body for the past 4 days, fever for 1 day, diffuse abdominal pain for 7 days with 3 episodes of vomiting on most of the days and severe arthralgia which made her unable to stand and walk. The patient also had multiple erythematous papules on both legs. It leads to the diagnosis of Henoch-schonlein-purpura. Proteinuria in this patient is a renal involvement of Henoch-schonlein-purpura. Symptomatic treatment was provided for this patient with paracetamol; an NSAID and prednisolone; a corticosteroid. Prednisolone is the most commonly used steroid in the treatment of HSP. Paracetamol as an analgesic had played a key role in alleviating fever and arthralgia associated with arthritis. Ondansetron was prescribed for vomiting and oral rehydration salt for dehydration which developed as a complication of vomiting.

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## INTRODUCTION:

Henoch-schonlein-purpura (HSP) is otherwise called immunoglobulin-A vasculitis [1]. It is the most common form of small vessel vasculitis. It affects the vasculature of several systems including joints, kidneys, gastro-intestinal tract and skin which results in multi-organ dysfunctional involvement [2]. Cutaneous manifestations are common in more than 90 % of paediatric cases. Antigen-antibody complexes, mostly of Ig-A deposition in the small vessel walls as a result of bacterial or viral infections, drugs, biologics, auto-immunity is the etio-pathogenesis of HSP. It is characterised by the presence of vasculitic purplish rash, abdominal pain, joint pain, vomiting, proteinuria [3]. Arthralgia and arthritis are

common characteristics of HSP in both paediatrics and adults [4]. About 3 to 27 cases per 100,000 children were encountered in this IgAV [5]. Older children, mainly the teenagers are affected from this IgAV which reflects the adult onset of IgAV [6]. The main goal of treatment for this disease is to provide acute symptomatic relief and prevent renal deterioration. Management of cutaneous involvement is usually not an absolute necessity [7]. This study discusses a clinical case of a teenage girl diagnosed with HSP. She also had one of the two signs of renal involvement i.e. proteinuria. Long-term follow is obligatory for the renal involvement of this disease [8]. The conditions like pott's spine and juvenile idiopathic arthritis are yet to be ruled out. Symptomatic treatment was given for symptoms like fever and vomiting. Prednisolone was used as a mainstay therapy for HSP.

## CASE REPORT:

### Description:

A 12 year old female child was admitted to the paediatric department of the Virudhunagar government medical college hospital with the complaints of red raised stain lesions over the body for the past 4 days, fever for 1 day, vomiting and diffuse abdominal pain for 7 days with 3 episodes of vomiting on most of the days and severe arthralgia which made her unable to stand and walk. The patient also had multiple erythematous papules on both legs. The patient's representative explained that she had suffered from similar conditions 1 year back and received treatment in a private setting. The patient also had complaints of back pain, pain over the lumbar region, and spinal tenderness. Under physical examination, the patient also showed positive kernig sign (a sign which is most commonly associated with meningitis).

### Diagnosis:

Recent diagnosis reveals that the patient has been suffering from Henoch Scholein Purpura and small vessel vasculitis. The patient was under evaluation for juvenile idiopathic arthritis and pott's spine.

### Investigation:

On investigating vitals, blood pressure was 100/60 mmHg, pulse rate was 88 beats/min, $SpO_2$ was 98 % and respiratory rate was 22 cycles/min.

On investigation of laboratory parameters, the total count was 33,000 cells/cubic mm. Under differential count polymorphs was 93 %, lymphocytes was 5 %, monocytes was 2 %. Red blood cells were 6 million/cubic mm, hemoglobin was 12.2 g/dL, packed cell volume was 43.5 % platelet count was 4 lakhs/cubic mm, erythrocyte sedimentation rate was found to be 42 mm/h. Bleeding and clotting time was found to be 2 and 5 min respectively. Renal function test reveals urea level as 38 mg/dl and creatinine level as 0.7 mg/dl. Random blood sugar level was 138 mg/dL. C-Reactive protein level was found to be 38 mg/L. Liver function test report reveals serum glutamate oxaloacetate transaminase concentration as 21 IU/L, serum glutamate pyruvate transaminase concentration as 20 IU/L, alkaline phosphatase concentration as 78 IU/L. Urine analysis reveals presence of albumin indicating progress of proteinuria.

Imaging study report of USG Abdomen reflects mild splenomegaly.

### Management:

The patient was under hospitalization for 5 days where she was treated with cephalosporin and aminoglycoside antibiotics, corticosteroid, antiemetics, antipyretic, and vitamin supplements.

The patient received normal saline and dextrose normal saline at a rate of 65ml/hr. Cefotaxime 1g by intravenous route twice daily for 5 days and amikacin 75 mg by intravenous route once daily for 5 days. Prednisolone (1 mg/kg/day) was administered by intravenous route thrice daily for 4 days. Ranitidine 25 mg was administered by intravenous route two times daily for 5 days. Ondansetron 2.5 mg was administered intravenously three times daily. About 500 mg of paracetamol was administered orally thrice daily for 5 days. She was advised to drink 125 ml of oral rehydration salt solution after every vomiting episode. Liquid paraffin ointment was prescribed to soothe skin rashes.

On discharge, the patient was advised to take tablet paracetamol 500 mg every $12^{th}$ hourly and ranitidine 25mg every $12^{th}$ hourly for 5 days, tablet vitamin-B complex 30.5 mg and tablet calcium 300 mg once daily for 15 days. Tablet prednisolone 5mg was advised to be taken every $12^{th}$ hourly for 15 days. She was advised for referral after 15 days to the outpatient setting of the same hospital.

## DISCUSSION:

HSP is an acute IgA mediated, leukocytoclastic vasculitis manifested as purpura (without thrombocytopenia), abdominal pain, and arthritis [9]. Its median age of onset is six years [10]. It is the most common vasculitis in paediatrics. Men are more prone to

HSP when compared to women [11]. This is a clinical case of a 12 year old adolescent girl diagnosed with HSP which is also not associated with thrombocytopenia. European League Against Rheumatism (EuLAR) and Pediatric Rheumatology Society (PReS) – 2006 developed criteria for the diagnosis of HSP [12]. The mandatory criterion is palpable purpura. Additional criteria are diffuse abdominal pain, Ig-A deposition in any biopsy, arthritis, and renal involvement (proteinuria and/or haematuria). The confirmatory diagnosis needs mandatory criterion plus one among the additional criteria. This clinical case has purpura in addition with diffuse abdominal pain, arthritis, and renal involvement (proteinuria) which fulfilled the diagnosis of HSA.

Common symptoms will be subsided except that of renal involvement. In this case also, symptoms like fever, arthralgia, purpura started to subside on the fourth day of hospitalization. Symptomatic treatment was provided for this patient with paracetamol; an NSAID, and prednisolone; a corticosteroid. Prednisolone is the most commonly used steroid in the treatment of HSP[13]. Paracetamol as an analgesic had played a key role in alleviating fever and arthralgia associated with arthritis. Ondansetron was prescribed for vomiting and ORS for dehydration which developed as a complication of vomiting.

The renal involvement of this disease has a high morbidity and mortality. This patient had only proteinuria but not haematuria. This renal pathology is also a self-limiting one in most of the cases. Around 7 % of patients suffered from nephritic condition and only around 1 % developed end-stage renal disease [14]. ACE inhibitors are found to be effective in the treatment of proteinuria associated with HSP [15]. It is recommended for this patient in order to attain a positive prognostic outcome.

## CONCLUSION:

This Henoch-schonlein purpura case of a paediatric patient with renal involvement needs referral to paediatric nephrologist to evade further complications. Other symptoms associated with HSP commenced to subside due to early intervention and treatment with steroid and paracetamol. This case study upholds the need to consider HSP in a broader epidemiological profile because some of them may have symptoms which are atypical in nature.

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