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Henoch-Schonleinpurpura with Sickle Cell Disease- A Rare Presentation

KEYWORDS: Homozygous sickle cell disease, sickle cell b - thalassaemia,hemoglobin,electrophoresis, Henoch-Schonleinpurpura,rash,renal disease.

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ABSTRACT *Henoch-Schonlein purpura (HSP) also known as Anaphylactoidpurpura or purpura rheumatic,is a disease of skin and other organs. Disease presents with palpable purpura with Joint, Kidney and abdominal involvement.*

HSP is a type of systemic vasculitis that means inflammation of blood vessels caused by deposition of immune complexes containing the IgA antibody, the exact cause is not known. It is self limiting disease, usually resolves within several weeks without treatment.

Systemic steroid are recommended for moderate to severe HSP. Steroid therapies lead to complete resolution of the symptoms.

Sickle cell disease is hereditary blood disorder associated with abnormality in the oxygen carrying hemoglobin molecule in red blood cells. This lead to abnormal shape of red blood cell that is rigid, sickle like shape associated with haemolysis, severe infection, pain (sickle cell crisis) and stroke. Sickle cell disease is common in tribal people of central India. The prevalence has ranged from 9.4 to 22.2% in endemic areas of Madhya Pradesh, Rajasthan and Chhattisgarh.

Introduction

Henoch Schonlein Purpura (HSP) is the commonest vasculitis seen in the pediatric population [1], with an incidence of 10-20 per 100, 000 children. Although treatment with corticosteroids is routine during the acute phase of the disease, controversy remains as to the merit of corticosteroids in preventing persistent renal disease in children presenting with HSP. Although several sporadic case series have claim demonstration of efficacious usage of prednisone and immuomodulators in preventing development of HSP nephropathies, data from randomized controlled trials are limited.

Sickle-cell disease is a multisystem disease, associated with episodes of acute illness and progressive organ damage, and is one of the most common severe monogenic disorders worldwide.[1] Herrick[2] first described the characteristic sickle-shaped erythrocytes in 1910 and understanding has gradually increased . Pauling and colleagues[5] identified electrophoretic abnormalities in sickle haemoglobin (HbS) and coined the term "molecular disease" in 1949. The haemoglobin biophysics and genetics underlying the disease have been extensively studied and have helped the understanding of other molecular diseases. However, clinical management of sickle-cell disease is still basic and, although some evidence lends support to the use of blood transfusion and hydroxeurea in some circumstances, no drugs have been developed that specifically target the pathophysiology of this disease.

We are reporting a case of Henoch–Schonlein Purpura (HSP) in a known case of sickle cell disease, we could not find any description of this entity on most popular online medical databases.**CASE REPORT:**

A 9 year old female from Alirajpur (MP) presented with **fever, rash** and **pain** in abdomen since 2.5 years. The rash was distributed all over all extremities including buttocks. Rash was erythematosus, non pruritic type which used to resolve spontaneously in 2-3 days (figure 1, 2, 3). Pain abdomen was colicky and intermittent type over left hypochondrium sometimes which was associated with 2-3 episodes of hematemesis.The patient also had associated history of **convulsion (Generalized Tonic Clonic type)** 3-4 episodes, each episode lasted for 4-5 minutes, resolved spontaneously without any medication. Patient had similar complain since past for that she had been hospitalized 3-4 times. On investigation haemoglobin 8.8 gm/dl, total leukocyte count 18900/cumm, Differential 64% neutrophils 26% lymphocyte, platelet counts 5.3 lacs/cumm, Prothrombin time of INR 1, ESR-33mm at 1sthr, C-reactive Protein 54.083 mg/L. On hemoglobin electrophoresis it was found to be sickle cell disease, Skin biopsy for Direct immunofluorescence was consistent with IgA vasculitis (figure 4, 5), Stool for Occult blood was positive, Urine examination was positive with albumin +2 on Dip stick and pus cells 5-6/hpf, epithelial cells 2-3/hpf. EEG shows mild degree of nonspecific diffuse electro physiological disturbance which was maximum over the right hemisphere. USG abdomen-Suggestive of lymphadenitis with sign of colitis.

Volume : 5 | Issue : 10 | October 2015 | ISSN - 2249-555X

Figure 1 : clinical picture of palpable purpura involving upper extremities

Figure 2: clinical picture of palpablepurpura involving lower extremities

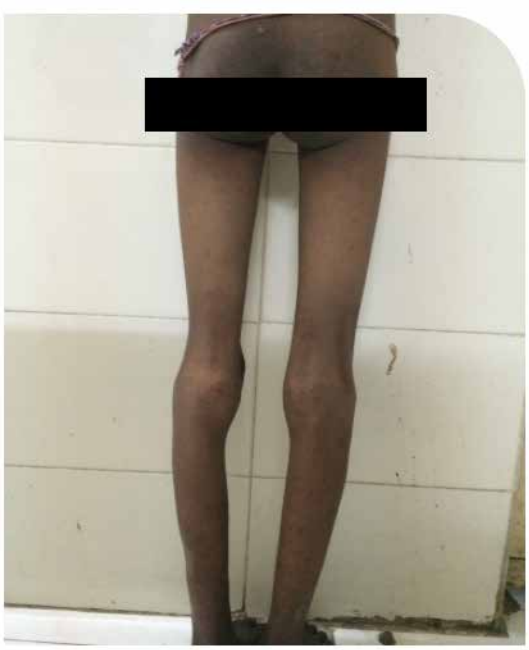

Figure 3:clinical picture of palpable purpura involvement of buttocks

Figure 4: IgA Vasculitis

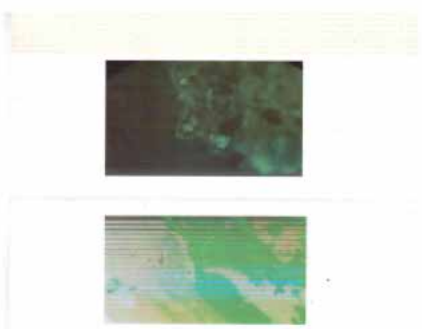

Figure 5: Granular positivity in papillary dermal vessels

DISCUSSION

Henoch-Schonlein purpura is characterized as self limiting systemic, non-granulomatous, small vessel vasculitis with multi-organ involvement.[3,5] It is most common cutaneous vasculitis in children less than 17 year of age with slight male predominance ratio (1.2:1 M:F). Incidence more at age of less than 10 year. It affects small arteries and veins due to antigen and antibody complexes mostly IgA from bacterial and viral infection or autoimmune mediated antigen antibody complex deposit in small vessel wall and activate alternate complement pathway which lead to neutrophil deposition resulting in vasculitis and inflammation occurs without a granulomatous reaction.[4]

CLINICAL FEATURES:

Cutaneous involvement is most common presentation although patient may present with involvement of multiple organs.

Skin:pink macules or wheels developing into petechiae, raised purpura or ecchymosis. The lesions are usually symmetric and occur in gravity dependent or pressure point(buttocks/lower extremities/distal end of upper extremities) and are <1cm initially but latter coagulase. subcutaneous edema to dorsum of hand/feets/periorbital area/lips/scalp are also common.[4 6]

Volume : 5 | Issue : 10 | October 2015 | ISSN - 2249-555X

Gastrointestinal: Abdominal pain (colicky in nature) is most common gastrointestinal symptom. Vomiting/ diarrhea/ paralytic ileus/ malena is also sometimes associated. GIT symptoms secondary to vasculitis (mesenteric ischaemia)/Intussusception(ileo-ileal)/perforation are rarely seen.

Renal: Haematuria is most common renal presentation which may or may not be associated with proteinuria. Proteinuria may also present alone though rare. Usually HSP nephritis is self resolving appear 1-3 wk after upper respitatory tract infection gross hematuria is seen in 20-30% of cases with microscopic hematuria, hematuria and proteinuria acute nephritic syndrome, nephrotic syndrome, are less common HSP nephritis occur up to 12 wk after presentation of HSP.[7]

Neurological: CNS involvement though very rare but may present with convulsion/intracerebralhaemorrhage/headache/behavioural change.

Joints:one third patients have joint pain as presenting symptom. Most commonly involved joints are ankle joint and knee joint.

Since our patient belongs to tribal area so we performed sickling test which was positive and to confirm whether patient has trait or disease Hb Electrophoresis was sent. Reports confirmed patient has sickle cell disease

Diagnostic Criteria(American College of Rheumatology) 2 of the following:

1. Palpable purpura
2. Age at onset<20yr
3. Bowel angina(postprandial abdomen pain, bloody diarrhea)
4. Biopsy demonstrating intramural granulocytes in small arterioles and/or venules.

European League against Rheumatism/Pediatric Rheumatology:
Palpable purpura(in absence of coagulopathy or thrombocytopenia) and one or more of the following criteria must be present:

1. Diffuse abdominal pain
2. Arthritis or arthralgia
3. Biopsy of affected tissue demonstrating predominant immunoglobulin-A deposition.[8]

TREATMENT:
Inj 0.45%DNS
Inj. Cefotaxime 1g
Injciplox (200/100)
Inj. Tramadol 50 mg
Inj. Phenytoin 50 mg
Inj. Vancomycin500mg
Inj. Levitracetam100mg
Tab. Prednisolone 20mg
Tab Valproate 200mg
Tab Cyclopam 100mg
Tab. Folc acid 5mg
Tab Brufeen 400mg
Cap. Hydra 500mg
Syp. Rantac

Conclusion: There are several challenges in treating a case of HSP with Sickle Cell Disease as there are no studies available online and steroid can be harmful to a patient of Sickle cell disease but as It is required in HSP. Because of the rarity of the presentation we are presenting this case report.

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