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Henoch Scholien Purpura: Not Just a Pediatric Diagnosis

Dr. Ankit Manglunia, Dr. Piyush Kumar, Dr. Vineet Kumar, Dr. Saumya Mathur, Dr. Aashish Sharma, Dr. Prakhar Garg, Dr. Vimal Mathur, Dr. GN Saxena

Department Of Medicine, MGUMST, Jaipur

ABSTRACT *: Henoch Schonlein Purpura (HSP) is a systemic vasculitis mediated by IgA and characterised by the clinical triad of non-thrombocytopenic palpable purpura, abdominal pain and arthritis. In addition, there may be varying degree of renal involvement. It is the most frequent vasculitis in children and the incidence in adults varies.*

CASE REPORT

39 year old male presented with history of recurrent renal stones since last 8-10 years. He underwent PCNL right side for the same in January 2015. Platelet counts were also 81000/uL during the same time. He developed non blanchable rash on the extremities and on lower abdomen and trunk a week later. The skin lesions were non pruritic and the lesions were both palpable and non-palpable. Patient developed left knee swelling associated with pain followed by right knee swelling and pain 3 days later and then the patient was admitted in our hospital. Laboratory Investigations showed HB – 14.2 gm/dL, WBC – 11.65*103 /uL with Neutrophils 66.4%, Eosinoplils- 1.1%, platelet count were 1.5 lakhs .Renal function tests and serum electrolytes were within normal range. Liver Function Test revealed slightly raised levels of SGOT/SGPT [89/187] with normal bilirubin level. Urine analysis was normal. ANA, C3 C4 levels was normal.Ig-A levels was 391mg/dl**.**Skin Biopsy was taken which showed focal upper dermal perivascular neutrophils with mild leucocytoclasis and some extravasated RBCs. Deeper sections shows similar features. Immunofluorescence stains show granular fluorescence of IgA in the papillary dermal vessels. Findings was suggestive of Small Vessel Vasculitis.

Figure 1: Skin Lesion of the Patient on Abdomen

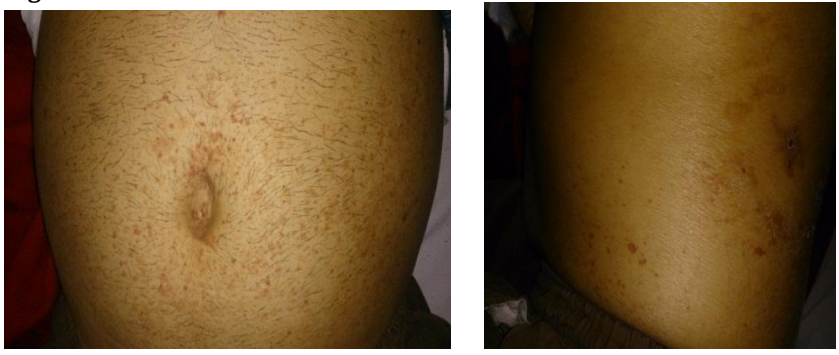
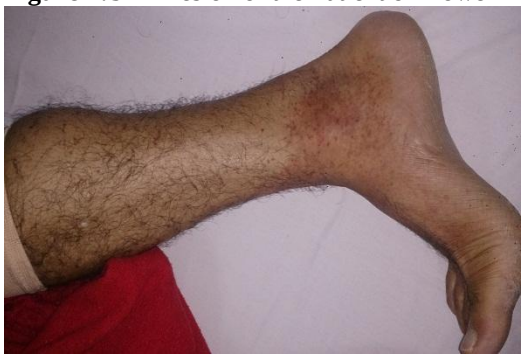


Figure 2: Skin Lesion of the Patient on Lower Extremity



DISCUSSION

HSP is a small vessel vasculitis characterised by IgA complexes deposition in tissues. It generally has a self-limiting course, and its main clinical manifestations can be seen in skin, the gastrointestinal tract, and the kidneys. Less frequently other organs and/or system, such as lungs, the CNS and the genito-urinary tract can be affected.

Though the etiology of the disease is unknown, multiple infectious agents have been suggested to be responsible. In adults, renal involvement and the possibility of progression to renal failure is greater as compared to children. The prevalence is greater in males.

Although there are no randomised control trials that prove the efficacy of one therapy over another (especially for kidney involvement), corticosteroids have been used at high doses, either orally or in pulses of methylprednisolone, and in association with immunosuppressive agents, such as cyclophosphamide or azathioprine.

Although HSP is infrequent in adults, renal involvement deserves special attention because it darkens the prognosis. Thus these patients should be detected and treated as early as possible.

CONCLUSION

Henoch-Scholien purpura is a fairly common paediatric disorder. The prognosis for the majority of patients is very good. Less than 2% of patients develop a serious complication associated with long-term morbidity. The most common serious complication is end-stage kidney disease, which may develop late in the disease after the symptoms from other organ systems have resolved. Patients with HSP should be carefully followed for several years to look for evidence of late renal involvement, and those patients who develop renal involvement should be followed regularly throughout their lifetime. Primary care physicians should be well aware of the disease because the true incidence is probably underestimated.

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