Case Report

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Henoch Schonlein Purpura

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Abstract

HSP is the most common childhood vasculitis, and is characterized by the classic tetrad of nonthrombocytopenic palpable purpura, arthritis or arthralgias, gastrointestinal and renal involvement. It is a systemic disease where antigen-antibody (IgA) complexes activate the alternate complement pathway, resulting in inflammation and small vessel vasculitis. Mild disease resolves spontaneously, and symptomatic treatment alone is sufficient. Systemic steroids are recommended for moderate to severe HSP. The prognosis depends upon the extent of renal involvement, which requires close followup. Early recognition of multiorgan involvement, especially outside of the typical age group, and appropriate intervention can mitigate the disease and limit organ damage. We describe a case of 6 year male with Henoch-Schonlein purpura (HSP), presenting with cutaneous and gastrointestinal manifestations

Keywords: Henochscolienpurpura; Palpable Purpura; Arthritis.

Introduction

Henoch-Schonlein purpura (HSP) is a self-limited, systemic, nongranulomatous, autoimmune complex, small vessel vasculitis, with multiorgan involvement. Its etiology is unclear but is associated with infections (bacterial, viral, parasitic), medications, vaccination, tumors (non-small cell lung cancer, prostate cancer, and hematological malignancies), alpha-1-antitrypsin deficiency, and Familial Mediteranean Fever [1].

HSP is the most common cutaneous vasculitis in children comprising up to 90% of cases [2,3]. Peak age incidence is 4–6 years and 90% of HSP cases occur before the age of 10 years. HSP is most commonly seen in winter and spring seasons. In adults, the incidence varies between 3.4–14.3 per million population. As this disease is self limited, its true incidence may be underreported [4, 5].

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Case Report

A 6 year male came with complaint of erythematous, nonpruritic rash which progressed proximally from both feet to thighs and upper extremities including palms and soles, patient also complaint of abdominal pain and joint pain 4 days before admission. Patient had been to local practioner, but symptoms didnt releived .

On physical examination, there was, a nodular, and nontender, nonblanching purpuric rash involving both upper and lower extremities. There was no truncal involvement. Two days later, the patient developed abdominal pain involving the right and left upper quadrant which was constant, colicky in nature, aggravated with meals.

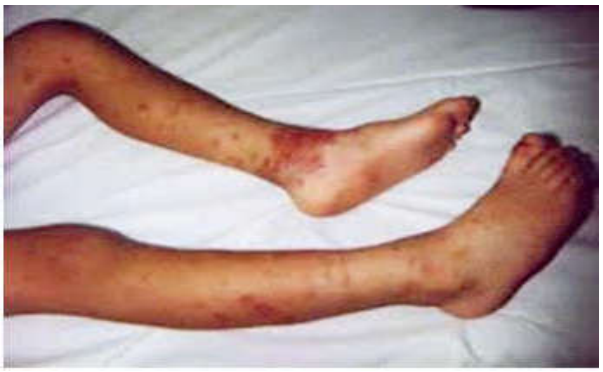

Fig. 1: Rash of Henoch schonlien Purpura

Laboratory Tests Showed

Leukocytosis (WBC: 16,900/microL);
Hb: 14 g/dL; Hct: 41.2%;
BUN: 16 mg/dL;
Serum Creatinine: 0.9 mg/dL;
Urinalysis: no hematuria or proteinuria;
ESR: 30 mm/Hr;
CRP: 5.6 mg/dL;
Antistreptolysin O titer: 150IU/L;
C3: 125 mg/dL;
C4: 11 mg/dL;
ANA: Negative; IgM:
Negative;
Anti-HAV IgM: Negative;
HbsAg: Negative;
Anti-HBc IgG: Negative;
Mono spot test: Negative;
c-ANCA: 0.2 units;
p-ANCA: 0.2 units;
Stool for occult blood: Negative.

Histopathology of the small bowel showed preserved villous architecture, and neutrophilic and eosinophilic infiltrates with leukocytoclastic vasculitis.

Treatment

He was treated with intravenous fluids. Intraveneous ISO -P was given on maintainance basis. He was started on oral prednisone 20mg twice a day, with resolution of his symptoms and a decrease in ESR and CRP. Patient was also given oral analgesic. Diet was a given regular diet during its hospital stay.

Discussion

Although HSP is uncommon in the adolescent age group, non-thrombocytopenic palpable purpura with multiorgan involvement (gastrointestinal, kidney and joints) should make one consider the diagnosis. Vasculitis can cause extravasation of blood and its components into the interstitial spaces which can result in edema and hemorrhage Prompt diagnosis and multidisciplinary intervention can lead to appropriate management and mitigate potential complications.HSP is mostly a clinical diagnosis but when the presentation is atypical, tissue biopsy may be helpful [6,7]. HSP is mostly self limiting. It carries a good prognosis. The five year survival rate been 95% [8,9].

Conclusion

A 6 year male coming to our paediatric OPD , complaining of nonpruritic rash which progressed proximally from both feet to thighs and upper extremities and also complaining of Abdominal pain and also joint pain was admitted . Patient was then treated with with intraveneous fluid ,analgesics and steroids. The disease is mostly self limiting ,diagnosis is mostly clinical.

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