

SEVERE GASTROINTESTINAL PRESENTATION OF HENoch-SCHÖNLEIN PURPURA WITH DECREASED FACTOR XIII ACTIVITY: A CASE REPORT

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Introduction: Henoch-Schönlein purpura (HSP) is a non-granulomatous, immunoglobulin A-mediated small vessel vasculitis primarily affecting children and young adults. It is characterized by its diverse clinical presentation, often encompassing cutaneous, joint, and renal symptoms. However, a subset of HSP cases exhibits a unique manifestation involving the gastrointestinal system. Here, we report a case of HSP with severe gastrointestinal manifestation.

Case report: A 5-year-old boy, previously healthy, presented two weeks before admission with a purpuric skin rash on his lower limbs. After two weeks, his condition deteriorated, and he was admitted, with new skin changes emerging on his upper limbs, genital and gluteal regions, accompanied by swelling in the joints, genital area, and transient microhematuria. He received oral prednisolone treatment, followed by parenteral prednisolone and a proton pump inhibitor. On the 7th day, he experienced severe episodes of abdominal pain accompanied by gastrointestinal bleeding (hematochezia) requiring a blood transfusion. Laboratory and imaging data revealed normal findings, except for the low level of factor XIII of 36% (normal range 50-150%). Cryoprecipitate transfusion was given twice, which shortly resulted in a cessation of abdominal pain and gastrointestinal bleeding, but the skin rash still persisted.

Conclusion: The convergence of gastrointestinal symptoms in Henoch-Schönlein purpura, along with cryoprecipitate transfusion for low factor XIII levels, presents a complex treatment scenario. Modifying approaches to match these specific details is essential for achieving improved patient outcomes.

Keywords: Henoch-Schönlein purpura; gastrointestinal manifestation; factor XIII; cryoprecipitate