

Abdominal Pain, Dysentery and Bilious Vomiting: An Atypical Presentation of Henoch Schonlein Purpura: A Case Study

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Abstract

Adolescent male resident of Hazara, Pakistan presented with abdominal pain, bilious vomiting, dysentery and knee pain of 10 days duration in July of 2014 to the emergency department of the Aga Khan University Hospital. Knee pain later migrated to ankle and was associated with redness and oedema. Abdominal x-ray revealed thickened bowel wall that was suggestive of enteritis. He was being treated with IV antibiotics (ceftriaxone and metronidazole), which did not help in resolving his symptoms. Overnight he started developing palpable rashes on his feet, back and abdomen, which raised the suspicion of Henoch-Schonlein purpura. There were no signs to suggest any respiratory and urinary systems involvement. He was given intravenous prednisolone after which his rashes dramatically disappeared and his ankle pain and other symptoms resolved. On follow up his lab investigations were within normal range with complete disappearance of rash and resolution of his abdominal pain, dysentery and bilious vomiting. Rarely dysentery is accompanied with palpable rashes and should raise suspicion of Henoch-Schonlein Purpura. Knowledge and understanding of this rare disease can help early diagnosis and initiate prompt treatment.

Keywords: Adolescent, vomiting, edema, enteritis, Henoch-Schonlein Purpura. (ASH & KMDC 20(1):61;2015).

Introduction

Henoch-Schonlein Purpura is the most common type of small vessel vasculitis and develops in children due to the deposition of immunoglobulin A (IgA) immune complexes in the small vessels walls leading to their eventual necrosis. Triggers for developing this condition include infectious agents, vaccinations and drugs. It mostly manifests as an acute onset palpable purpura in the lower extremities¹. The disease can virtually involve any organ system of the body and therefore can present with a wide range of symptoms, these may include abdominal

pain, diarrhea, dysentery, subcutaneous edema, scrotal edema, joint pain, renal involvement etc. A detailed physical examination is therefore indicated. It is a disease of young children with 50% below 6 years and 90% younger than 10 years. It rarely develops in adolescent and adult population². Here we present a case of an adolescent male patient with an atypical presentation of Henoch-Schonlein Purpura that was misdiagnosed initially and resulted in a prolonged hospital stay.

Case Report

An 18 year old male patient presented in the emergency department of The Aga Khan University Hospital Karachi with history of loose stools 4-5 episodes per day which he explained to be watery in consistency containing small amounts of fresh blood. He had central abdominal pain moderate to severe in intensity, colicky in nature with mild vague

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Fig. 1.1. Palpable purpuric rashes visible on the back



Fig. 1.2. Confluent pattern of rash observable over the dorsum of bilateral feet



Fig. 2. X-ray abdomen supine view showing prominent thick walled small bowel loops abdomen

tenderness on examination. There was no association with meals and body movements. He also complained of 3-4 episodes of non-projectile vomiting, greenish with food particles and small amount of blood in it. He developed bilateral knee pain, which later migrated to both the ankles. Later he also developed swelling at both the ankles, which caused difficulty in walking. He was afebrile throughout and had no urinary symptoms. Our patient had never experienced such symptoms in the past neither he had any history of allergies, blood transfusions and any significant family history of bleeding disorders, malignancies or autoimmune diseases. On examination he was alert, active and fully oriented pale looking young individual. There was no appreciable jaundice, cyanosis or clubbing however he was mildly tachycardic with a pulse of around 100 with systolic pressure of around 110 without any fever. On neurological examination, he had no focal deficits; power was 5/5 in all four limbs, though movement at both knee and ankle joints were painful (7 on scale of 10).

Cardiovascular examination was unremarkable, S1 and S2 audible with no added sounds without any murmurs, thrill or heave. Respiratory system examination revealed normal vesicular breathing bilaterally without any crackle, wheezing or rhonchi on auscultation. Abdomen was soft, non-distended but tender in mid epigastrium only. Gut sounds were clearly audible. No surgical marks, bruises or scars were noted. Among lab investigations Blood Urea Nitrogen, Creatinine and electrolytes were sent to check for any electrolyte abnormalities and status of volume depletion. Complete Blood Count was sent to assess the Hematocrit level for indirect assessment of blood loss, TLC counts for assessment of acute or chronic inflammatory response and platelet counts for normal clotting function. Our patient had normal hematocrit of around 30 with raised TLC counts of 13000. All other laboratory investigations were within normal range. Abdominal X-ray was done in order to rule out any mechanical cause of intestinal obstruction, X-ray revealed: Non-specific bowel gas pattern. However the small bowel folds appeared thickened (Fig. 2). Findings

were suggestive of gastroenteritis. Differentials were bacterial or viral acute gastroenteritis, mesenteric adenitis and intestinal intussusception. He was given IV fluids along with metronidazole and ceftriaxone keeping bacterial gastroenteritis as provisional diagnosis but his pain and other symptoms did not resolve. On 2nd day of his hospital stay he developed purpuric rashes on his feet, buttocks, and lower back Fig.1.1 which were red, raised, palpable and non-tender, and had confluence pattern Fig.1.2.

Our clinical diagnosis was confirmed by skin biopsy of the rash taken from dorsum of right foot. The histopathology revealed classical picture of Henoch Schonlein Purpura with leukocytoclastic vasculitis involving the post capillary venules with IgA deposition. His treatment was started on clinical suspicion prior to availability of biopsy report with intravenous prednisolone 40 mg administered twice daily. Miraculously his abdominal pain, dysentery and rashes dramatically improved including his ankle joint pain and swelling. He was afebrile throughout the hospital course and was discharged on tapering dose of oral steroids.

At his follow up after a week in outpatient clinic his white counts had reduced to normal with complete resolution of rashes over the body, diarrhea, vomiting and joint aches.

Discussion

Henoch-Schönlein purpura (HSP) primarily affects children with an annual incidence of 13-20 cases per 100,000 <17 years old children³. However data regarding its prevalence in Pakistan is not available due to lack of data reporting and registries with limited occurrence of the disease itself. The rare nature of the disease, its self-resolving pattern, and limited resources of histopathology to confirm the diagnosis could explain difficulty in diagnosis. Henoch Schonlein purpura is a systemic vasculitis with multiorgan involvement. According to the European League Against Rheumatism, presence of Purpura or petechiae is a mandatory criterion for diagnosis of HSP⁴.

However in our case diffuse abdominal pain and acute joint pain were present with no petechiae or purpura, which did not raise our suspicion of an autoimmune etiology. Our initial treatment was focused towards an infectious etiology, which had led to gastro intestinal disturbance and blood in stool. However with the appearance of palpable purpuric rashes we started considering about a vasculitic source of abdominal pain with diarrhea and thus a suspicion of Henoch Schonlein Purpura were raised. Amongst the atypical presentations of HSP one case was reported in which the patient presented with status epilepticus with delayed development of rash over two weeks⁵. Local studies from our region have reported an unusual case of HSP in which a 40 year old was diagnosed with *Helicobacter pylori* infection by upper GI Endoscopy while he presented with upper abdominal pain, retrosternal burning and palpable purpura over the lower limbs⁶.

A similar case was reported in which patient presented with abdominal pain mimicking that of appendicitis and Appendectomy was performed. He was diagnosed later as a case of HSP when his pain did not resolve post-operatively and he developed purpuric rash on lower extremity⁷.

The delayed appearance of rash is very uncommon in HSP patients and in our case this resulted in delay in reaching a final diagnosis. So abdominal pain and acute onset of joint pain in a young patient should raise suspicion of vasculitis syndrome

Corticosteroids are effective on arthralgias and abdominal pain, but ineffective on skin purpura, and there is a considerable controversy on the benefit of corticosteroids to treat renal involvement and prevent evolution to end-stage renal disease. Other recent treatment options include Colchicines, Dapsone, anti-leukotriene agents, Azathioprine, Myophenolatemofetil, Cyclosporin A, Cyclophosphamide and Rituximab depending upon the degree of involvement of organs systems⁸. A meta-analysis by Weiss et al took into account various studies regarding effects of corticosteroids in Henoch Schonlein purpura, Heterogeneity was noted in the

resolution of abdominal pain after administration of steroids which was likely due to timing of starting corticosteroids and it was concluded that Corticosteroid treatment did not reduce the median time to resolution of abdominal pain but did significantly reduce the mean resolution time⁹. A 3 year clinical analysis of 254 cases of HSP recommended that steroid treatment given to HSP patients with gastrointestinal involvement is helpful to prevent complications including GI bleeding and intussusception; however combined therapy of steroids with cyclophosphamide (or cyclosporine if resistant to cyclophosphamide) can be used for patients with nephritic proteinuria¹⁰. However more intensive therapy with methylprednisolone pulse therapy should be limited to patients at risk of progression to severe nephrotic syndrome and/or crescent glomerulonephritis as discussed in a randomized double blind placebo-controlled trial¹¹.

The case just reported was unlikely to be suspected as a case of HSP due to the age of the patient and delayed appearance of rash. Initial gastrointestinal symptoms were misleading and did not respond to antibiotics and fluid resuscitation. Later with the development of rash suspicion of an autoimmune cause was raised with biopsy proving Henoch Schonlein Purpura. Our patient responded to corticosteroid therapy with cessation of GI bleeding episodes and resolution of diarrhea.

Conclusion

Rashes are mandatory criterion for the diagnosis of Henoch Schonlein purpura but it might appear late in the course of the disease which could lead to misdiagnosis, inappropriate treatment and prolonged hospital stay. One should have a high degree of suspicion in patients presenting with dysentery not responding to antibiotic therapy for Henoch Schonlein Purpura. Early steroid therapy in Henoch Schonlein Purpura has shown various beneficial effects in the resolution of abdominal pain, rashes and other gastrointestinal symptoms; however its role in prevention and treatment of renal involvement is not yet established.

References

1. Torrelo A, Noguera L. Henoch-Schönlein Purpura. In: Silverberg NB, Duran-McKinster C, Tay YK, editors. *Pediatric Skin of Color*. New York: Springer; 2015. p. 417-9.
2. Iemsawatdikul K, Daldrup-Link HE. Henoch-Schönlein purpura. In: Daldrup-Link HE, Newman B. *Pearls and Pitfalls in Pediatric Imaging: Variants and Other Difficult Diagnoses*. Cambridge: Cambridge University Press; 2014. p. 205.
3. Yang YH, Tsai IJ, Chang CJ, Chuang YH, Hsu HY, Chiang BL. The interaction between circulating complement proteins and cutaneous microvascular endothelial cells in the development of childhood Henoch-Schönlein Purpura [Internet]. *PLoS One* 2015;10:e0120411. Available from: <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC4356510/>
4. Ozen S, Pistorio A, Iusan SM, Bakkaloglu A, Herlin T, Brik R, et al. EULAR/PRINTO/PRES criteria for Henoch-Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part II: final classification criteria. *Ann Rheumatic Dis* 2010;69:798-806.
5. Mannenbach MS, Reed AM, Moir C. Atypical presentation of Henoch-Schönlein purpura. *Pediatr Emerg Care* 2009;25:513-5.
6. Shaikh MZ, Ahmed A. Henoch-Schönlein purpura associated with gastric helicobacter pylori infection. *Pak J Med Sci* 2004;20:68-70.
7. Hernstadt HM, Bartlett M, Kausman JY, Macgregor D, Akikusa JD. Complicated Hench-Schönlein purpura. *J Paediatr Child Health* 2014.
8. Audemard-Verger A, Pillebout E, Guillevin L, Thervet E, Terrier B. IgA vasculitis (Henoch-Schönlein purpura) in adults: Diagnostic and therapeutic aspects. *Autoimmun Rev* 2015;14:579-85.
9. Weiss PF, Feinstein JA, Luan X, Burnham JM, Feudtner C. Effects of corticosteroid on Henoch-Schönlein purpura: a systematic review. *Pediatrics* 2007;120:1079-87.
10. Peru H, Soylemezoglu O, Bakkaloglu SA, Elmas S, Bozkaya D, Elmaci AM, et al. Henoch Schonlein purpura in childhood: clinical analysis of 254 cases over a 3-year period. *Clin Rheumatol* 2008;27:1087-92.
11. Dudley J, Smith G, Llewelyn-Edwards A, Bayliss K, Pike K, Tizard J, et al. Randomised, double-blind, placebo-controlled trial to determine whether steroids reduce the incidence and severity of nephropathy in Henoch-Schonlein Purpura (HSP). *Arch Dis Child* 2013;98:756-63.