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CASE REPORT

HENOCH-SCHONLEIN PURPURA: A TYPICAL PRESENTATION

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ABSTRACT: Henoch-Schonlein Purpura (HSP) is the most common vasculitis of childhood and is characterized by leukocytoclastic vasculitis and immunoglobulin (Ig) A deposition in the small vessels in the skin, joints, gastrointestinal tract, and kidney. It is a systemic disease where antigen-antibody (IgA) complexes activate the alternate complement pathway, resulting in inflammation and small vessel vasculitis. Here we are reporting an interesting case of an adolescent female with atypical presentation (hypovolemic shock, erosive gastritis, skin involvement of the extremities) requiring very high degree of suspicion for the diagnosis.

KEYWORDS: Hematemesis, Erosive Gastritis, Cholecystitis, HSP, Henoch-Schnolein Purpura, Palpable Purpura.

CASE: A 13 years old female patient was referred to our hospital with complaints of fever, pain abdomen, joint pains and rashes over lower-limbs since 3-4 days duration and 7-8 episodes of emesis of which 4-5 episodes were frank blood since 1 day.

Patient when received in the ER was in hypovolemic shock. Other examination included, child had multiple Petechial rashes over her B/L wrists.

A provisional diagnosis of Viral Hemorrhagic Fever with shock was proposed.
Patient was immediately resuscitated with IV fluid boluses and hemodynamically improved.

Her Complete Blood Counts were normal and there was evidence of Thrombocytopenia in the peripheral smear. Clotting Profile (PT, a PTT, INR, BT and CT) were all within normal limits for age, Dengue serology was negative and an USG abdomen revealed features of Cholelithiasis with free fluid. Patient had further episodes of hematemesis, an upper GI endoscopy revealed Gastric Erosion.

Child was started on IVF's, antacids, PPI, Sucralfate and NPO.

During the course of PICU stay, the rashes seemed to disappear, hematemesis subsided but the patient continued to have severe pain abdomen, colicky in nature, with tenderness in the Right subcostal and epigastric region. A surgery consultation was taken and a Contrast CT abdomen was ordered which revealed multiple tiny Cholelithiasis with minimal free fluid in the para-colic gutter. Patient was continued on symptomatic treatment and her general condition improved over a period of time and was started on soft oral diet which was tolerated well.

On day 8 of hospital stay, the patient again developed new rashes which were purpuric in nature, palpable, present over both lower limbs and buttocks [fig]. Perilesional skin biopsies for immunofloresence revealed IgA deposition and C3 deposition, suggestive of Henoch-Schonlein Purpura.

Urine Routine and Kidney function test were within normal limits. Child was started on oral steroids. Rashes gradually disappeared.

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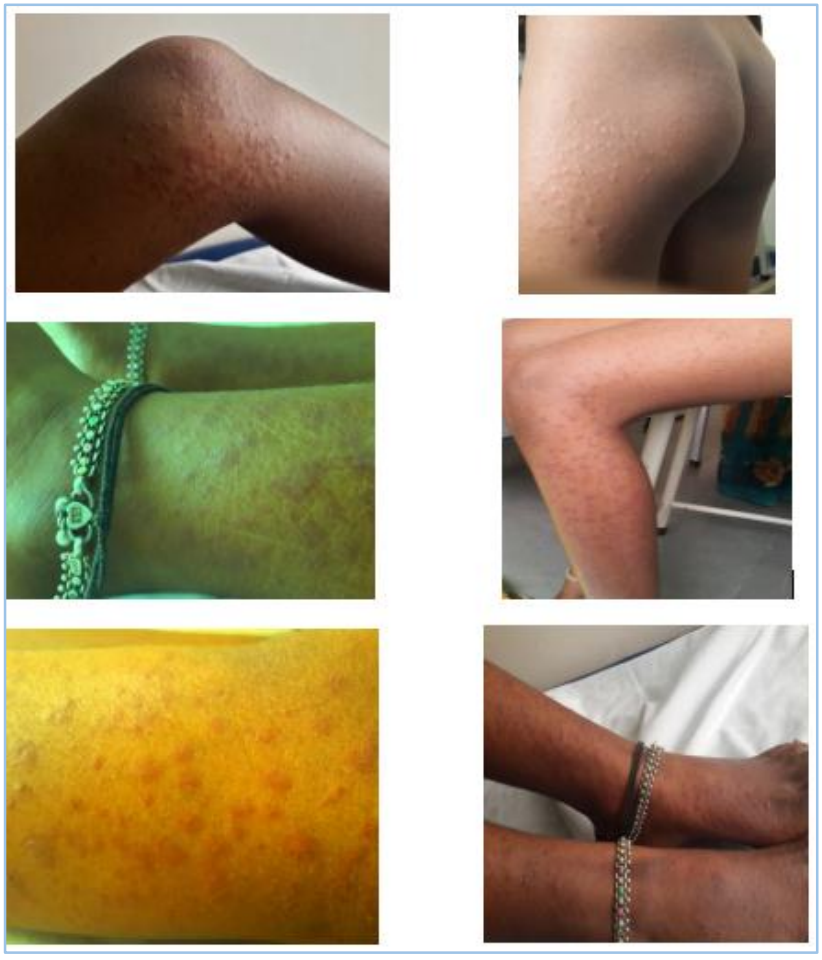
Figures Showing Palpable Purpurae over the Gravity Dependant Areas:



DISCUSSION: HSP is a form of blood vessel inflammation or vasculitis, also referred to as anaphylactoid purpura.[1] Clinically, Henoch–Schonlein purpura presents with a classic tetrad of symptoms (with reported frequencies): cutaneous purpura (100%), arthralgia or arthritis (82%), abdominal pain (63%), gastrointestinal bleeding (33%).[1] A biopsy of the skin, and less commonly, the kidneys, can be used to demonstrate the presence of vasculitis.[2] Special staining techniques (Direct immune-fluorescence) of the biopsy specimen can be used to document antibody deposits of IgA in the blood vessel or tissue.[2]

According to the diagnostic criteria of the European League against Rheumatism and the Pediatric Rheumatology European Society published in 2006, palpable purpura often presents with one of the following: diffuse abdominal pain, a biopsy showing predominant IgA expression, acute arthritis/arthralgia, or renal involvement defined as any hematuria or proteinuria.[3]

However, when gastrointestinal manifestations occur alone or precede dermatological or renal disease, the diagnosis is difficult.[1] the clinical manifestations of HSP in the gastrointestinal tract are similar to gastrointestinal tuberculosis.[2] lymphoma, inflammatory bowel disease, and other autoimmune disorders, which renders the diagnosis of HSP difficult. In this case, drastic abdominal pain with hematemesis[4] and hypovolemic shock[5] was presented as the princeps clinical situation.

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The characteristic endoscopic findings included patchy gastric erosions.[6,7] but no petechiae or hemorrhages. These manifestations along with cholecystitis[8] created confusion until the diagnosis of HSP was verified by the presence of IgA deposition in perilesional skin biopsies.[9]

CONCLUSION: HSP can present as any other viral hemorrhagic fever when only gastrointestinal symptoms are present without typical palpable purpuric rashes. During course of illness it can be complicated with cholecystitis and erosive gastritis, the cause for hematemesis in our case. The incidence of hematemesis is known to be around 13.1% as per the literature.[4] Palpable purpuric rashes with typical distribution can appear very late during course of illness, sometimes when the patient has completely recovered from symptoms as in our case.

Cholecystits, erosive gastritis, thrombocytopenia during initial course of illness and late appearance of typical palpable purpuric rashes delayed our diagnosis of HSP in this case.

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