Photo Quiz

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A 4.5-year-old boy with Diffuse Purpuric Lesions all Over his Extremities

A 4.5-year old boy is admitted to the pediatric Emergency Room with diffuse purpuric lesions all over his extremities. He is alert but has a fever (38.4 ^{o}C). He also has slight joint swelling in the left knee and both ankles.

He had a fever and sore throat for the past two days. His primary care doctor gave him oral penicillin. From today morning, he developed the rash which progressed proximally from both feet to thighs and upper extremities including palms and soles.

His past medical history is unrevealing. Family history is negative for any significant disease. The review of systems is unremarkable. The mother says she has not given any medication to him. On physical examination, he is a tachycardic boy with a blood pressure of 130/90 mmHg and a respiratory rate of 22/min. Eyes are normal, the soft palate has several petechiae and the face shows mild edema. Both hands are mildly edematous. There are diffuse non tender, nonblanching purpuric lesions on the arms, legs, and buttocks (Fig 1). The lungs and heart examination are normal. The abdomen is diffusely painful on palpation without rebound tenderness or guarding and without liver and spleen enlargement.

His laboratory results showed: Hemoglobin: 11.9 g/dL (reference, 11-14), WBC count:19.8 × 10^3/µL (reference, 6-14), platelets: 510 × 10^3/µL (reference,150-450), C-reactive protein:63.5 mg/L (reference, <0.5), ESR: 56 mm/hr, blood urea nitrogen:32 mg/dL (reference, 12-42), serum creatinine: 0.48 mg/dL (reference, 0.32-0.59), total protein: 49 g/L (reference, 62-78), albumin:30 g/L (reference, 35-52), C3: 125 mg/dL(reference, 80-160), C4: 11 mg/dL(reference, 16-48), ANA: Negative, c-ANCA: 0.2 units/mL (reference, ≤3.5), p-ANCA: 0.2 units/mL (reference, <0.4), electrolytes, coagulation times, liver enzymes, and lipase were within normal range.

Urinalysis: nephrotic range proteinuria (3.2 g/L; reference, 0-0.2), positive hematuria, positive leukocytes (370/µL; reference, 0-25). Abdominal ultrasound showed no abnormalities of liver, kidneys, or spleen and adequate peristalsis of the intestines.

What is your diagnosis?

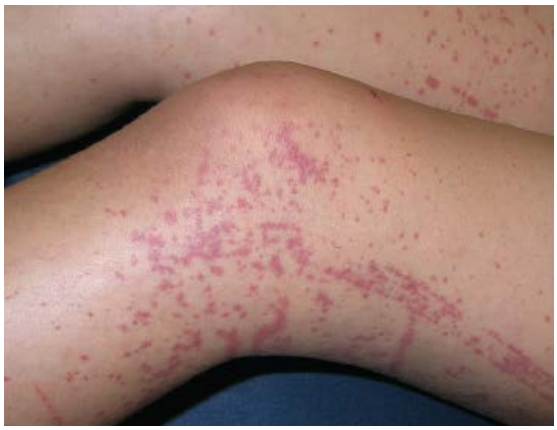

Figure 1. Diffuse nontender, nonblanching purpuric lesions on the left knee.

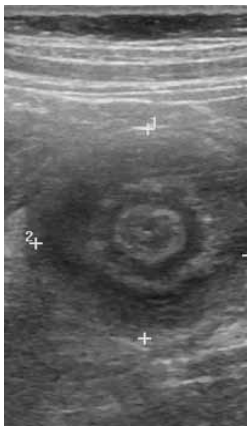

Figure 2. Target Sign in abdominal Ultrasound, showing abnormal cecum

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Q1: What is the most probable diagnosis for him?

A1: Based on the American College of Rheumatology and European League Against Rheumatism (EuLAR) and Pediatric Rheumatology Society (PReS) criteria, he is diagnosed with Henoch-Schonlein purpura.

Q2: What is the best treatment for him?

A2: IV fluid, High-dose pulse IV steroid.
Symptomatic treatment is usually enough for symptoms such as rash and arthritis. Acetaminophen and NSAIDs can be used. Aspirin must be avoided in children.

Oral steroids are indicated in patients with severe rash, edema, severe colicky abdominal pain (without nausea, vomiting), renal, scrotal, and testicular involvement. Usually, prednisone or methylprednisolone can be started at 1 to 2 mg/kg per day for one to two weeks, tapering down to 0.5 mg/kg/day over the next week and then 0.5 mg/kg every other day for one more week. Intravenous (IV) steroids can be administered if the patient does not tolerate oral steroids.

High-dose IV pulse steroids are indicated in patients with nephrotic range proteinuria and mesenteric vasculitis. These pulse doses can range from 500 mg to 1 gm with various protocols leading to complete remission of nephritis (1,2).
Two days later, he develops diffuse abdominal pain with bloody stools (4-6 times a day). An enlarged and painful left scrotum is seen. Blood and urine cultures are negative. Ultrasound of the scrotum showed diffuse enlargement and hypervascularization of left epididymis consistent with epididymitis.

Q3: What is the reason for bloody stool?

A3: Small and large intestine petechial involvement (3)

Q4: What is the best treatment for his epididymitis?

A4: Continuing the steroid treatment (1)
Despite the treatment, hematochezia persists for the next 48 hours. Then, he suddenly develops severe colicky abdominal pain with reduced peristalsis. On physical examination, an elongated mass is detected in the right upper quadrant with an absence of bowel in the right lower quadrant (Dance's sign). The abdominal radiograph shows ileus. The abdominal ultrasound examination shows a "target sign" (Figure 2), which is due to the cecum in the periphery and the intussuscepted

small intestine in the middle, while no subdiaphragmatic free gas is seen on the abdominal radiograph.

Q5: What is the diagnosis for him now?

A5: ileocecal intussusception

Q6: What is the best treatment for him?

A6: Air enema

Patients with intussusception should be assessed for the presence of peritonitis and the severity of systemic illness. Following resuscitation and administration of IV antibiotics, the child is assessed for suitability to proceed with radiographic versus surgical reduction. In the absence of peritonitis, the child should undergo radiographic reduction. If peritonitis is present, or if the child appears systemically ill, urgent laparotomy is indicated (4).

In a stable patient, the air enema is both diagnostic and may be curative, and it is the preferred method of diagnosis and treatment of intussusception. Air is introduced with a manometer, and the pressure that is administered is carefully monitored. In most instances, this should not exceed 120 mmHg. The successful reduction is marked by free reflux of air into multiple loops of the small bowel and symptomatic improvement as the infant suddenly becomes pain-free. Unless both of these signs are observed, it cannot be assumed that the intussusception is reduced. If the reduction is unsuccessful, and the infant remains stable, the infant should be brought back to the radiology suite for a repeat attempt at a reduction after a few hours. This strategy has improved the success rate of non-operative reduction in many centers. In addition, hydrostatic reduction with barium may be useful if the pneumatic reduction is unsuccessful. The overall success rate of radiographic reduction varies based on the experience of the center, and it is typically between 60% and 90% (4).

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