A 14-Year-Old Boy with Bullous Lesions

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A 14-year-old previously healthy boy presents with a 2-week history of purpuric rash with worsening pain of lower extremities. He has an unremarkable past medical and surgical history, with age-appropriate development. Two weeks prior to presentation, he developed “bug bites” around his ankles bilaterally with minimal pruritis. Over the next 3 days, this rash began to progress to his mid-calves and coalesce into larger purplish lesions. One week prior to admission, he began to have severe abdominal pain forcing him into the fetal position. There was no associated nausea or vomiting with the abdominal pain; however, the episodes of pain would be intense and then gradually resolve. There is no history of fever or joint pain. Family history is noncontributory, and there is no recent travel or sick contacts. There are no known allergies.

PHYSICAL EXAMINATION

He is alert and oriented in mild discomfort due to the pain in his lower extremities. His vital signs are: temperature, 36.1°C; pulse 64 beats/min; blood pressure 103/56; and respiratory rate, 14 breaths/min. There is no conjunctivitis, no cervical lymphadenopathy, or pharyngeal erythema. Lung and cardiovascular exam are within normal limits. He has a soft, non-distended abdomen with normal bowel sounds, no organomegaly, and no guarding. An integumentary exam revealed numerous purplish purpura with multiple blisters and hemorrhagic bullae on the lower extremities surrounding the ankles and extending to the dorsum of his feet and up his calves bilaterally (see Figure 1, right). There is mild edema of the ankles bilaterally (see Figure 2, page 276). His neurologic exam is within normal limits.

LABORATORY FINDINGS

Laboratory examination revealed a total white blood cell count of 7 x 10^3/μL, with 57% polymorph nuclear cells, no bands, 30% lymphocytes, and 4% eosinophils. Hemoglobin was 14.4 g/dL, Hematocrit was 39.1%, and platelets 357 K/μL. Coagulation studies revealed normal prothrombin time. The basic metabolic panel was within normal limits, with normal blood urea nitrogen (BUN) and creatine. The urinalysis was negative and stool was negative for occult blood.

For diagnosis, see page 276

Editor’s note: Each month, this department features a discussion of an unusual diagnosis in genetics, radiology, or dermatology. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at pediatrics@healio.com.
Diagnosis:
Henoch-Schonlein Purpura

Henoch-Schonlein purpura (HSP) is a common systemic vasculitis in children. This disease primarily occurs between the ages of 3 and 15 years with an estimated annual incidence of 20 per 100,000 children, with the highest incidence between ages 4 to 6 years (70 per 100,000 children). HSP is an immune-mediated vasculitis associated with IgA deposition, and while a variety of infectious triggers have been speculated, the underlying etiology remains unknown.

Classically, HSP affects skin, gastrointestinal tract, joints, and kidney. However, the order of their presentation may vary, and the disease may progress over days to weeks. Patients may present with only joint pain and/or abdominal pain, without the classic purpuric rash, which may initially raise concern for a surgical or infectious process.

• **Skin manifestations**: The rash often begins as erythematous macules that coalesce into petechiae and palpable purpura. The rash is often symmetric in distribution and located in pressure-dependent areas such as the lower extremities and buttocks. Bullous presentation as seen in this patient is not common, but is a recognized cutaneous manifestation that may make initial diagnosis difficult. Unlike adult HSP, hemorrhagic bullous evolution has been rarely described in children. Fortunately, a review of three pediatric patients with severe hemorrhagic bullous lesions did not indicate a poorer prognosis of HSP or related complications.

• **Gastrointestinal symptoms**: Gastrointestinal symptoms may range from nausea, vomiting, or abdominal pain to a more severe presentation of bloody stools, intussusception, and bowel perforation. Guaiac-positive stool is found in up to 56% of patients, but massive gastrointestinal hemorrhage is rare. Intussusception is the most common gastrointestinal complications of HSP, and is generally limited to small bowel so ultrasonography can be the initial screening test.

• **Arthritis/arthralgia**: The arthritis may be transient or migratory, and often involves lower extremity large joints and less commonly upper extremity joints. There is often joint swelling with pain and limited range of motion, but without joint effusion, erythema, or warmth.

• **Renal disease**: Manifestations of renal disease range from isolated hematuria and/or proteinuria to nephropathy with renal insufficiency. In a retrospective review, renal involvement was found in approximately 52% patients, ranging from microscopic hematuria to nephritic syndrome.

Diagnosis of HSP is based on clinical findings and no laboratory test is diagnostic. Normal platelet count and coagulation studies are necessary to exclude other disease presentations. Urinalysis should be performed on all patients with suspected HSP to evaluate the degree of renal involvement. However, for patients with atypical presentation, biopsy may demonstrate leukocytoclastic vasculitis with a predominance of IgA deposition.

DISCUSSION

Management of HSP generally focuses on supportive care, yet there are indications for targeted therapies to decrease risk of complications. Supportive care focuses on adequate hydration and pain relief. Patients with severe or prolonged abdominal pain may require...
intravenous hydration or parenteral nutrition. Some patients may develop abdominal obstruction and require surgical evaluation. Patients with significant bleeding may require red blood cell transfusion; patients with renal involvement who are hypertensive may require medications for blood pressure control. Severe joint pain or arthritis may be relieved by acetaminophen or nonsteroidal anti-inflammatory drugs (NSAIDs).

The use of glucocorticoids in HSP remains controversial. Reported benefits of glucocorticoid therapy in HSP has included shortening the duration of abdominal pain, decreasing the risk of intussusception, decreasing HSP recurrence risk, and decreasing the risk of renal involvement. A randomized-controlled trial to evaluate the efficacy of prednisone therapy found that steroids reduced severity of abdominal pain and joint pain, and were effective in altering (but not preventing) renal manifestations. A meta-analysis by Weiss et al attempted to answer the question of whether glucocorticoids are beneficial. Their analysis concluded that glucocorticosteroids may shorten the duration of abdominal pain and risk of developing persistent renal disease; however, there were inherent limitations to the study in that steroid regimens varied and definition of renal involvement varied.

CONCLUSION
The use of systemic steroids (1mg/kg/day) has been described for patients with bullous lesions in HSP to reduce the severity of HSP related bullae and its associated painful ulcers and necrosis. This patient was started on steroids (1 mg/kg/day divided BID) and his pain improved within 24 hours. The bullous lesions improved gradually over several weeks, and his renal function was monitored carefully with frequent blood pressure checks and urinalysis. His steroids were gradually tapered over 4 weeks, and he did not develop any subsequent renal or gastrointestinal complications. ■

REFERENCES
Target Audience
This conference is designed for all pediatric professionals – office pediatricians, pediatric nurse practitioners, family practitioners and other primary health care providers.

Educational Objectives
At the conclusion of this activity, attendees should be able to:

- Summarize the American Academy of Pediatrics Committee on Infectious Diseases and Centers for Disease Control and Prevention Advisory Committee on Immunization Practices recommendations for key vaccinations in the pediatric practice and recent improvements in vaccinations.
- Discuss periodic fevers with parents of children affected by Periodic fever, Aphthous-stomatitis, Pharyngitis, Adenitis (PFAPA) syndrome.
- Differentiate between fact and falsehood regarding Lyme disease.
- Describe neonatal viral infections and the most appropriate treatments.
- Apply evidence-based guidelines for pediatric care of infectious diseases to everyday clinical practice.
- Evaluate pneumonia in children and differentiate between viral and bacterial causes.
- Incorporate new technologies and treatments, including immunomodulatory treatments, in cases of Kawasaki disease.
- Evaluate various skin disorders and determine the best method of treatment.
- List the key features of the latest antimicrobial agents.
- Incorporate current guidelines and evidence for the prevention and management of resurgent measles.
- Summarize the current protocols and guidelines regarding the treatment of animal and insect bites.
- Differentiate and treat syphilis in the pediatric population.
- Describe clinical symptoms for human papillomavirus (HPV) and apply preventative measures.
- Evaluate currently available antibiotics and use them judiciously.
- Summarize the epidemiology of lymphadenopathy and evidence-based treatment and prevention strategies.
- Diagnose and manage sinusitis.
- Apply the latest developments in the Staphylococcus aureus epidemic to daily practice.
- Manage patients with rotavirus in the most efficient and effective manner possible.

NEW FORMAT
To achieve a well-balanced program, we’ll round up “The Usual Suspects” in our morning review sessions and update your practice with sessions on “The New Developments” in the afternoons.

SPOT THE RASH
This new addition to the meeting will bring the popular monthly challenge you see in print to a new level with interactivity. Presenters will test your skills on pediatric dermatology issues, kicking off the meeting with a fun and exciting mental workout.

TOP 10
To round out the meeting, our experts will carefully review journal articles published throughout the year and provide perspective and action items on the 10 most applicable to your daily practice.

CLASSROOM-STYLE
We have eliminated round tables and are implementing a classroom setup for improved visibility and to foster greater interaction between our experts and colleagues.
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