Jake's Achin' Abdomen

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Jake's presentation

- Jake, 23, previously healthy, presents to the emergency department with a dull, diffuse abdominal pain.
- The pain is associated with nausea and vomiting and loose stool, but no gastrointestinal blood.
- Jake denies any anorexia, fever, travel or other symptoms, but does complain of muscle pains for the same duration as the abdominal pain.
- Although he does not report it, you notice a red rash that, on exam, is palpable and confined to his lower extremities (see Figures 1 and 2).
- The remainder of Jake's physical exam is unremarkable—in particular, there are no meningeal signs and the abdomen is benign.

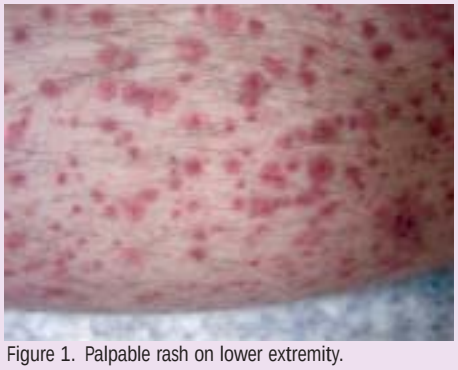

Figure 1. Palpable rash on lower extremity.

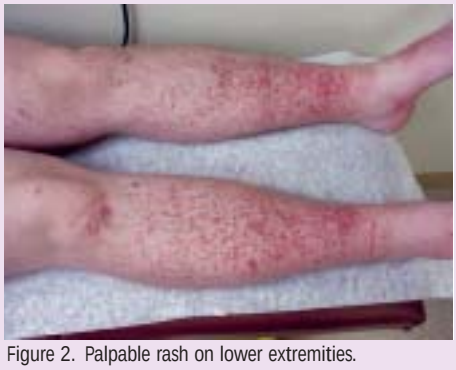

Figure 2. Palpable rash on lower extremities.

Questions & Answers

1. What is the diagnosis?

Henoch-Schonlein purpura (HSP) is likely. HSP is an immunologic mediated systemic vasculitis affecting small vessels. It is primarily a disease affecting children (majority of onset between age four and 11, with an annual incidence of 15/100,000), but adult cases can also be seen. The male/female ratio is 1.5-2.0:1. HSP has a variable clinical presentation that can involve the skin, gut, joints and kidneys. Most children present with purpura (Figure 1) localized to their lower limbs and buttocks and 70% have gastrointestinal (GI) complaints. These include colicky abdominal pain, nausea, vomiting and diarrhea or constipation that may include blood or mucus per rectum. Adults often present with skin lesions (Figure 2) and arthralgias. HSP is commonly preceded by an upper respiratory tract infection, with a peak incidence in the spring.[1-5]

2. How is the diagnosis confirmed?

The diagnosis is primarily clinical and, in otherwise healthy children, is based on a triad of palpable purpura, GI complaints and arthralgia (usually ankles and knees). Meningococcemia must be ruled out, especially if the presentation is accompanied by a fever, headache or a high white blood cell count.[5] Diagnosis may be more difficult in adults, especially when GI symptoms precede the rash. In one study of adults, a rash was found to be present in 95% of patients, arthralgia/arthritis (ankles and knees) in 60%, and GI symptoms in just under half (48%), with the main complaint being colicky pain.[2] A complete blood count should be performed, which often shows leukocytosis with an increase in eosinophils, as well as thrombocytosis. Urine analysis often shows hematuria and proteinuria, which are indicative of renal involvement. The erythrocyte sedimentation rate and serum immunoglobulin levels are usually increased. Severe and persistent abdominal pain may require a computerized tomography scan to rule out ileoileal intussusception in young children.

3. What major complications should be considered and how do they differ in children versus adults?

The most common complication of HSP is renal disease, often in the form of glomerulonephritis. Renal complications appear less in children (about 20% of the time[1] versus 32% of the time in adults).[3] Adult renal complications often present more insidiously, progressing through a number of years, whereas in children they present more acutely, usually within weeks of initial clinical presentation. Adult renal complications also tend to be more severe and, therefore, require close monitoring. Adult patients with renal complications often have persistent disease, clinically evident as hematuria and proteinuria, and remission is difficult to obtain, despite treatment. Also of note, children may suffer intussusception as a complication secondary to HSP gut involvement.

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4. What is the standard treatment?

Most HSP patients can be treated symptomatically. Children, who usually have less severe manifestations and lower incidences of complications, rarely need corticosteroid treatment. When they do, 1 mg/kg/day, with a 60-mg maximum, is effective in treating GI complaints and arthralgias.[5] Prednisone does not appear to have an effect on the skin rash, nor does it prevent renal complications, according to one randomized, controlled trial, although this remains controversial.[1] Similar benefits with steroids have been seen in adults; steroids are given more commonly because of their aggressive course in adults.[2] Cyclophosphamide is often used in combination with steroids in adult patients with renal complications; consultation with a nephrologist is usually warranted in these cases. Dx

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