March 2018 post: HSP (by Stan L. Block MD FAAP)

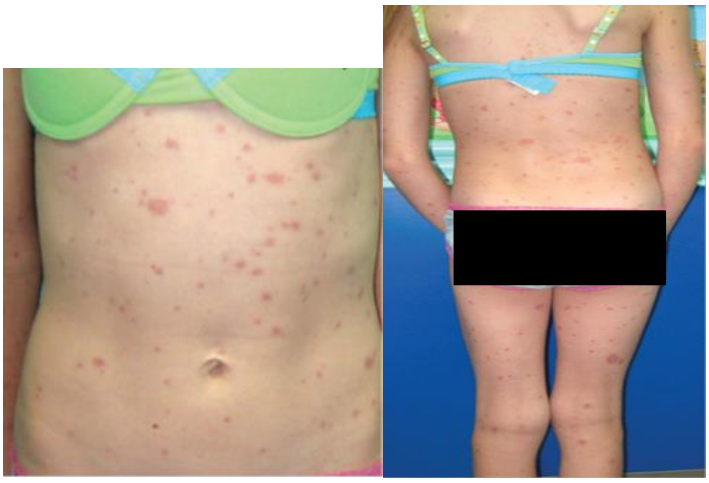
The great mimicker.

In mid-April, the pleasant otherwise healthy 16 yo WF presents to your office today with abrupt onset of multiple joint pains, most severe in her knees, wrists and elbows. She has been afebrile, and denies any sore throat (currently), vomiting, diarrhea, rash, cough, dysuria or vaginal discharge. She had a sore throat last month with some "fever" for 1.5 days, for which she did not seek medical care. Her family history is negative for any arthropathies or HLA B 27. Her examination is otherwise unremarkable, except for the exquisite joint pains over her knees and elbows, which you can barely examine or move them due to the pain. You do not appreciate any effusions in any joints. She needs assistance walking too.

You order your usual arthritis panel of tests, including ESR, CRP, CBC, ANA, RF, CMP, and titers for EBV, Lyme, and parvovirus. You have observed several teenage girls over the years who present with minimal or no dermatologic signs of Fifth disease, but who had moderate to severe joint pains and positive parvovirus titers, particularly in the late Spring months. Their mild flushed cheeks are often attributed to sunburn or windburn. You are also concerned about the joint pains having some migratory properties and being quite severe. That along with her undiagnosed sore throat last month, you are considering the possibility of acute rheumatic fever (ARF). Her ECG was normal. Since she is afebrile and looks well otherwise, you are not really concerned about Lyme disease, RMSF or meningococcemia, each of which can present with arthritis symptoms.

You send her home on ibuprofen three times a day, and ask her to return in 48 hours for re-evaluation. Her blood tests have all returned as normal by then, and her joint pains have only minimally eased up. No new signs or symptoms have cropped up, so you are beginning to think of more chronic arthropathies like JIA, SLE, Crohns, psoriaritic, or even undiagnosed Lyme, etc. You are now in a waiting game to see if she responds to ibuprofen, or if she develops any other physical clues, or if repeat testing of acute inflammatory markers and Lyme titers is needed. However, on day 5 she returns to the office with a very disturbing rash, nearly identical to the photographs of the patient below.

The following photographs and discussion are excerpted from an article that I wrote for *Pediatric Annals* in the August 2014 issue. The article also discusses the general evaluation of petechiae and purpura in the outpatient office setting. A different 13 y.o. WF with the typical rash of HSP, but with an atypical distribution, i.e., on the arms and entire back and abdomen.



This rash is termed "palpable purpura." "The distribution of the rash is too extensive to be typical for HSP or immunoglobulin A (IgA) vasculitis. **Or is it?** After all, you have been taught that the rash rarely goes superior to the waistline or on the arms. But one of the keys to this diagnosis of HSP is the usual lack of any systemic illness signs or fever." And yes, this rash is also HSP.

You are well aware that the IgA vasculitis (HSP) can primarily affect (early on or later) four other organ systems besides the skin, such as: 1) glomerulonephritis, 2) abdominal pain (rarely along with intussusception), 3) arthritis, and 4) orchitis/ oophoritis. Although these sequelae have been reported in 50% to 75% of children (who were mostly hospitalized), they are actually uncommon (< 5%-10%) in your experience with outpatients in the general pediatrics office. You are quite attuned to possible secondary nephritis, which may have particularly severe long-term sequelae and may require antihypertensive medication. Some of the patients may experience severe abdominal pain to the point of requiring surgical evaluation and hospitalization. If a patient is febrile despite an initial diagnosis of HSP and a typical HSP rash, it is important to obtain a blood culture. One of my febrile patients with HSP had a blood culture obtained in the emergency department which revealed that she actually had a meningococcal serogroup B infection. So be very careful with the diagnosis of HSP in **the febrile child**. A continued physical and laboratory evaluation is very important.

Monitoring in HSP

Careful follow-up over the first few weeks is still essential, despite the low incidence of nephritis. In the otherwise uncomplicated afebrile case after HSP diagnosis is made, you may typically recommend: 1) two additional visits within the first week, and then two more weekly visits; and 2) to return at any time if the family notices the child has developed puffy eyes, bad headaches, off-colored urine, decreased urine output, abdominal/genital/joint pain or swelling; 3) at each visit, a CBC, UA, and serum chemistries as well as obtaining weight, vital signs (blood pressure especially), and a physical examination.

Treatment of HSP

This is a fairly controversial area, but the literature suggests a possible role for steroid therapy, at least in hospitalized patients. I think that two recent studies conducted by Weiss et al, have shown that steroids may cause a modest reduction in renal disease and significant reductions in surgery (odds ratio: 0.39), endoscopy (odds ratio: 0.27), and abdominal imaging (odds ratio: 0.5).

When the child with HSP did not have severe abdominal pain, I have personally only used oral steroids in an outpatient setting one time. And, the results appeared to be dramatic within 24 hours of initiation of oral steroids. My rationale was based on the severity of his vasculitic rash in this case and the fact that steroids have also shown a modest benefit in some cases of severe mononucleosis, (which this child concomitantly had). The earlier steroids are started in more severe cases of HSP, the greater the benefit will be, in my opinion. Obviously, I did not want to wait for severity requiring hospitalization."

Reference: Block S, Petechiae and Purpura: The ominous and the not so obvious? Practical Advice for treating newborns and toddlers PEDIATRIC ANNALS • Vol. 43, No. 8, 2014

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