

Dermacase

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CAN YOU IDENTIFY THIS CONDITION?

An 11-year-old boy presented complaining that he had had a non-itchy rash on his legs and buttocks for 3 days. He had slight abdominal discomfort, but otherwise felt well. Examination revealed purpuric papules scattered over his posterior thighs. His right ankle was mildly edematous, but the rest of his examination was unremarkable.

The most likely diagnosis is:

1. Papular urticaria
2. Systemic lupus erythematosus
3. Henoch-Schönlein purpura
4. Meningococemia
5. Dermatitis herpetiformis

Answer on page 512

Dr Freiman is a dermatology resident at the McGill University Health Centre in Montreal, Que.

Answer to Dermacase *continued from page 511***3. Henoch-Schönlein purpura**

Henoch-Schönlein purpura (HSP) is one of the most common forms of vasculitis of childhood. It is characterized by immunoglobulin A deposits in the walls of small blood vessels.¹ The estimated annual incidence of HSP is 20.4/100 000 people and is highest in boys between the ages of 4 and 6.²

Clinically, HSP presents with a classic tetrad of symptoms that have documented relative frequencies: cutaneous purpura (100%), arthralgia or arthritis (82%), abdominal pain (63%), and gastrointestinal bleeding (33%).³ Characteristic primary skin lesions are purpuric papules symmetrically distributed on the buttocks, legs, and extensor extremities. Arthralgia commonly affects the knees and ankles, but resolves without permanent damage to joints. Gastrointestinal bleeding and intussusception are rarer complications of HSP and are presumably initiated by edematous vasculitis of the small-bowel mucosa.⁴ Occasionally, the central nervous system and the respiratory system are also affected.

The serious sequela of HSP is its renal involvement, which typically takes place within a few days to several weeks after onset of systemic symptoms. Heralded by asymptomatic microscopic hematuria and proteinuria, it occurs in 30% to 70% of patients, but it is usually mild and self-limiting. With increasing age at presentation, however, more marked complications can arise, including nephrotic syndrome, hypertension, and acute renal failure. Children diagnosed with HSP need their renal status closely monitored with repeat urinalyses and renal function tests and referral to a pediatric nephrologist if necessary.

The etiology of HSP is unclear. It is frequently associated with upper respiratory tract infection, which is consistent with its peak occurrence in the winter and fall. Patients usually present with a 2- to 3-week history of fever, headache, myalgia, arthralgia, and abdominal pain preceding the typical cutaneous purpura. Group A streptococci, mycoplasma,

and a variety of other infectious agents and drugs have been reported as potential triggers. Diagnosis of HSP is often made on the basis of clinical signs and symptoms and can be confirmed with direct immunofluorescence of the skin biopsy sample, which demonstrates leukocytoclastic vasculitis with perivascular immunoglobulin A, immunoglobulin C3, and fibrin deposits.

Henoch-Schönlein purpura is usually benign and resolves spontaneously, so treatment is mostly supportive, including adequate hydration. Long-term prognosis largely depends on the severity of renal involvement. About 94% of children and 89% of adults recover completely.⁵ Nonsteroidal anti-inflammatory drugs can be used to treat arthralgia associated with HSP. While systemic corticosteroids might help with some symptoms, such as arthritis and abdominal pain, no form of therapy has yet been shown in randomized trials to have an effect on the duration of illness or recurrences. Corticosteroids, cytotoxic agents, and intravenous immunoglobulin therapy might be of benefit in advanced disease, defined as crescentic nephritis.⁶

References

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