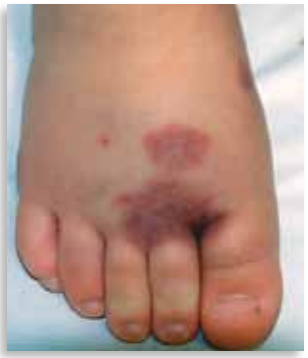
Answer to Dermacase *continued from page 666*

2. Acute hemorrhagic edema of infancy



First described in 1913 as the infantile variant of Henoch-Schönlein purpura (HSP), acute hemorrhagic edema of infancy (AHE) is now considered to have several characteristic clinical and prognostic features that qualify it as a distinct disorder.[1-4] Among these features are disease onset before the age of 2 years, peculiar skin manifestations, and rare visceral complications.[2]

Acute hemorrhagic edema of infancy is an acute leukocytoclastic vasculitis usually affecting children younger than 2 years of age, with a slight predominance in boys.[2,3] Symptoms are usually limited to the skin and manifest as strikingly large (ie, 0.5 to 4 cm in diameter) purpuric skin lesions in a targetlike pattern and marked edema of the face, ears, and extremities.[2-4] Although the disorder has an alarmingly dramatic acute onset, the course is usually benign and spontaneous resolution occurs within 1 to 3 weeks.[2] Extracutaneous involvement is rare, occurring in less than 10% of cases, and includes glomerulonephritis, abdominal pain, and arthralgia.[2] However, these tend to resolve completely with the resolution of the skin lesions.[2] Laboratory investigation results are usually within normal limits, although leukocytosis, eosinophilia, and a high erythrocyte sedimentation rate might be present.[2] Our patient had spontaneous resolution of the eruption within 10 days with no complications, and her laboratory results revealed only an elevated erythrocyte sedimentation rate.

The etiology of AHE is still not clear. It is regarded by many to be a hypersensitivity reaction, as most cases are usually associated with a history of recent infection (eg, upper respiratory tract infection, otitis, conjunctivitis, skin infection, infectious diarrhea, or urinary tract infection), drug intake, or immunization.[2,3] Its higher incidence in winter months also suggests an infectious etiology.[2]

Differential diagnosis

The most important differential diagnoses of AHE include HSP, erythema multiforme, and meningococcemia.[2,5] Although AHE is a separate clinical entity, it shares features with HSP, which indicates that both are closely related disorders within the spectrum of leukocytoclastic vasculitis.[2,3] Therefore, it is important to make a definite diagnosis of AHE from the standpoint of a prediction of a benign prognosis. Compared with AHE, HSP usually affects older children, shows smaller purpuric lesions, and is more commonly associated with systemic abnormalities. In addition, although both conditions share a similar histology, AHE lacks the immunoglobulin A deposition around small blood vessels, a direct immunofluorescence microscopy feature characteristic of HSP.

Erythema multiforme usually presents as a self-limited, acute, and occasionally recurrent skin eruption. It typically manifests as multiple classical target lesions, mainly over acral sites with minimal mucosal involvement. Large purpuric patches with associated edema are usually absent. Like AHE, erythema multiforme is also thought to be a hypersensitivity reaction as it commonly occurs after certain infections, such as herpes simplex virus and mycoplasma, or after taking certain medications (eg, nonsteroidal anti-inflammatory drugs, penicillins, and sulfonamides).[6] Unlike AHE, histology of erythema multiforme reveals acute interface dermatitis.

Patients with acute meningococcemia (caused by *Neisseria meningitidis*) are usually sick-looking and often exhibit fever, neck stiffness, and altered mental status, in addition to numerous small, gray-purple, stellate purpuric lesions (occasionally with slightly depressed centres and raised borders) in up to 90% of cases.[5]

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Competing interests
None declared

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