Diagnostic Challenge

A painful rash

Jeremy M. Rose, MD*; Ryan Sustrik, MD†; Shane Silver, MD‡; Anthony Herd, MD§

CASE HISTORY

An 80-year-old female resident of a nursing facility with a history of type 2 diabetes and dementia presented with a painful rash that began on her neck and subsequently spread to approximately 70% of her body. The rash began 1 month prior to her presentation. It involved the patient's palms and soles but was not present on her mucous membranes. The rash was initially pustular, but as it worsened, large areas of skin sloughed off (Figure 1), leaving tender erythematous sections without deep erosions (Figure 2). Erythematous plaques with dry white scale were visible on the patient's scalp. The patient was afebrile and had a pulse of 109 beats/min, a respiratory rate of 20 breaths/min, a blood pressure of 136/86 mm Hg, and a room air oxygen saturation of 96%. She had a white blood cell count of 27.8×10^9/L (91% neutrophils) and blood glucose of 7.3 mmol/L. The patient had received a course of oral prednisone, which had been tapered in the week preceding her presentation. Two months before coming to the emergency department, she had completed a course of terbinafine for an unrelated condition.

QUESTION

What is the likely diagnosis?

a) Staphylococcal scalded skin syndrome
b) Acute generalized exanthematous pustulosis
c) Acute generalized pustular psoriasis
d) Stevens-Johnson syndrome
e) Toxic epidermal necrolysis

For the answer to this challenge, see page 370.

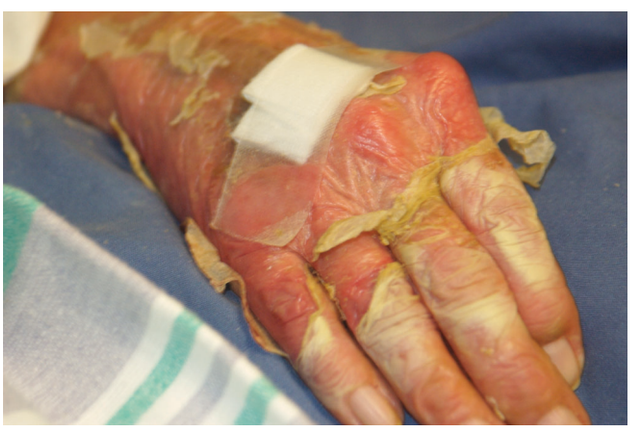

Figure 1. The patient's hand showing skin that had soughed off with exposed erythematous sections.

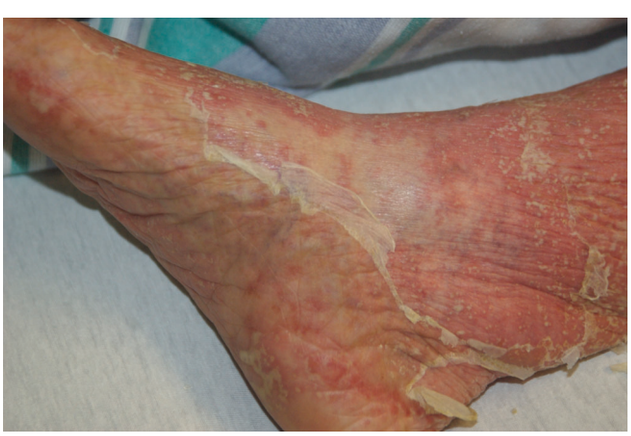

Figure 2. The patient's foot with a pustular rash.

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ANSWER

The correct answer is c) acute generalized pustular psoriasis. This is a form of psoriasis that is characterized by the formation of pustules rather than plaques. The lesions begin as large areas of erythema followed by clusters of sterile pustules. As the pustules grow, they merge. Superficial layers of skin may slough off, as in this case. Nikolsky sign is usually positive. The exposed layers are erythematous but do not feature deep erosions. Fever and leukocytosis are common. Treatment usually includes the use of oral retinoids. Although the etiology is unknown, generalized pustular psoriasis can be triggered by the cessation of oral steroids,[1] as was probably the case with this patient.

Pustular psoriasis is treated with acitretin (Soriatane). Notably, systemic steroids should be avoided as this can cause the disease to flare. Burn unit admission may be warranted for severe cases.

Our patient had previous psoriatic-type lesions and a history of steroid withdrawal, leading us to the diagnosis. The patient received treatment with acitretin and supportive skin care. Prednisone was not restarted. Skin biopsies revealed sterile subcorneal pustules containing neutrophils. The underlying dermis was edematous and contained an inflammatory infiltrate. The biopsy result was consistent with both psoriasis and acute generalized exanthematous pustulosis (AGEP). Unfortunately, the patient passed away 9 days after admission to hospital from complications of her illness.

How does this diagnosis differ from the other four differential diagnoses?

Staphylococcal scalded skin syndrome (SSSS) more commonly presents in children, although the mortality rate is much higher in adult patients. SSSS is the result of a toxin-mediated process. It may be preceded by a fever, sore throat, conjunctivitis, or other indication of an underlying infectious process and usually begins in flexural areas. SSSS causes generalized erythema and large, flaccid bullae, which can desquamate in areas of friction,[2] but it does not feature pustules. Although the patient in this case presented with a similar rash, she also had plaques consistent with psoriasis. More importantly, she had no focus for potential staphylococcal infection, making SSSS much less likely.

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are immune-mediated diseases characterized by necrosis of the epidermis. Both diseases are defined by the extent of the epidermal involvement. SJS generally affects less than 10% of body surface area, whereas TEN affects more than 30% of body surface area. Both diseases can arise from idiosyncratic drug reactions. Certain classes of drugs, such as antibiotics, nonsteroidal antiinflammatory drugs (NSAIDs), and anticonvulsants, are more commonly implicated in TEN. Allopurinol is a common culprit. SJS and TEN generally involve the mucous membranes, palms, and soles. The lesions initially appear as macules. As the macules grow, the epidermis begins to detach, forming fluid-filled bullae. Eventually, the epidermis necroses and sloughs, leaving deep erosions and exposing a tender, bleeding dermis.[3]

We initially suspected that our patient had TEN. However, on closer examination, her disease did not involve the full thickness of her epidermal layer.

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Furthermore, her mucous membranes were intact, and pustules are not a characteristic feature of TEN.

AGEP is a drug reaction that typically presents with an eruption of numerous sterile pustules days to weeks after exposure to the implicated medication. Classically, AGEP also presents with a high fever and leukocytosis. As the condition resolves, desquamation of the affected areas can occur.[4] Clinically and histologically, AGEP can be identical to pustular psoriasis. Our patient did have a remote history of terbinafine use, one of the drugs implicated in AGEP. Accordingly, we could not exclude AGEP from our differential diagnosis, although it was less likely given that the symptoms developed 1 month after cessation of that drug. AGEP has no specific treatment except for withdrawal of the causative agent and supportive care.

For the challenge, see page 369.

Competing interests: None declared.

Keywords:

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