Generalized Erythematous Plaques and Pustules in a Pregnant Patient

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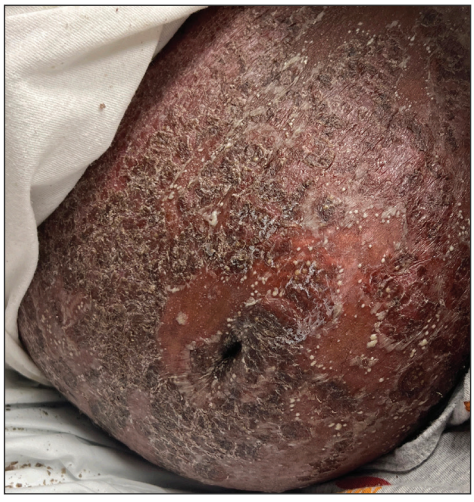

A 17-year-old girl was admitted to the hospital at 19 weeks' gestation for a widespread eruption of erythematous plaques with pustules covering more than 60% of the body and signs of sepsis. The rash initially appeared as a few small spots on the upper chest and under the breasts 5 weeks prior to hospital admission with subsequent spread to the abdomen and groin. At admission, the patient had a mild fever and tachycardia. She reported a history of eczema, herpes simplex virus, and intertrigo. Physical examination performed by dermatology revealed generalized erythematous plaques with pustule-studded margins and overlying scale involving the neck, torso, arms, and legs favoring the flexural areas. There was no involvement of the face, eyes, oral mucosa, palms, soles, or nails. Laboratory testing revealed hypoalbuminemia (2.4 g/dL [reference range, 3.5-5.5 g/dL]) and elevated inflammatory markers, including leukocytosis (15.83×10^3/μL [reference range, 4.50-11.00×10^3/μL]), absolute neutrophil count (12.87×10^3/μL [reference range, 1.50-8.00×10^3/μL]), and erythrocyte sedimentation rate (124 mm/h [reference range, 0-20 mm/h]). A culture from an abdominal pustule grew 1 colony of *Staphylococcus epidermidis*, a suspected contaminant. A biopsy from a lesion on the right chest was performed.

WHAT'S YOUR **DIAGNOSIS?**

a. acute generalized exanthematous pustulosis
b. dermatitis herpetiformis
c. impetigo herpetiformis
d. pemphigoid gestationis
e. subcorneal pustular dermatosis

PLEASE TURN TO PAGE 93 FOR THE DIAGNOSIS

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THE **DIAGNOSIS:**

Impetigo Herpetiformis

Histopathology revealed epidermal acanthosis and spongiosis with overlying parakeratosis associated with subcorneal and intracorneal neutrophils, papillary dermal edema, and dermal mixed inflammation with neutrophils and eosinophils. Both direct immunofluorescence and periodic acid–Schiff studies were negative. Blood and pustule cultures were sterile and the skin flora were normal. Based on these findings, a diagnosis of impetigo herpetiformis (IH) was made. The condition improved with systemic and topical steroids, supportive care, and an intravenous infusion of infliximab 5 mg/kg. At 3 weeks' follow-up, the patient demonstrated near-complete resolution and later delivered successfully at 40 weeks' gestation without complications.

Impetigo herpetiformis, also known as pustular psoriasis of pregnancy, is an exceedingly rare gestational dermatosis that typically manifests in the third trimester and can be life-threatening for both the mother and fetus. The term was first used in 1872 to describe 5 pregnant women with extensive acute pustular eruptions, all in unstable condition; 4 (80%)of the cases resulted in maternal death, and all resulted in fetal death.[1] Impetigo herpetiformis is characterized by pruritic and painful erythematous patches studded at the periphery with subcorneal pustules. Eruptions usually occur in the flexural areas and spread centrifugally, with extension of the lesions peripherally as the center erodes and crusts. Sparing of the face, palms, and soles is expected, and mucosal involvement is rare. Generalized involvement and exfoliation may occur in extreme cases.[2] While IH typically manifests during the third trimester, it may occur any time throughout pregnancy or immediately postpartum.[3] A few cases have been reported in the puerperium.[2] Common symptoms include fever, chills, malaise, anorexia, nausea, vomiting, diarrhea, and arthralgias. Less common complications include hypoalbuminemia and severe hypocalcemia leading to tetany, seizures, and delirium.[2,3] While maternal mortality is uncommon, fetal mortality often is a more pressing risk and is attributed to placental insufficiency.[3,4] For this reason, early delivery commonly is considered in severe cases.

Whether IH is a separate entity or a variant of pustular psoriasis remains heavily debated. Although the histopathology of IH is identical to pustular psoriasis, the lack of a personal and family history of psoriasis, symptom resolution with delivery, and possible recurrence during successive pregnancies help differentiate IH from generalized pustular psoriasis.[2,5] Earlier onset, diffuse involvement, faster progression, and recurrence in subsequent pregnancies all have been linked to a worse prognosis.[4]

The differential diagnosis for IH includes acute generalized exanthematous pustulosis, pemphigoid gestationis, dermatitis herpetiformis, and subcorneal pustular dermatosis. Acute generalized exanthematous pustulosis is an uncommon severe cutaneous drug reaction characterized by the sudden onset of numerous sterile pustules on erythematous skin within 48 hours of exposure. The most common offending medications include pristinamycin and beta-lactam antibiotics. A high fever, neutrophilic leukocytosis, and hypocalcemia often accompany acute generalized exanthematous pustulosis.[6] Prompt diagnosis and withdrawal of the offending drug as well as supportive care and symptomatic treatment are crucial for disease management, as systemic symptoms and even organ involvement may occur.[6]

Pemphigoid gestationis, also known as gestational pemphigoid or herpes gestationis, is a rare autoimmune blistering disorder that primarily affects pregnant women. It typically manifests in the second or third trimester or shortly after delivery. Clinically, it manifests as an intensely pruritic polymorphic eruption of urticarial papules and plaques accompanied by vesicles and bullae and often is distributed on the abdomen and extends to other body regions. Although the exact etiology is unknown, pemphigoid gestationis is caused by autoantibodies targeting the BP180 and BP230 hemidesmosomal proteins.[7] Treatment usually involves systemic corticosteroids and may require additional immunosuppressive therapy. In most cases, patients see resolution within 6 months of delivery.[7]

Dermatitis herpetiformis is a chronic autoimmune blistering skin disorder characterized by intensely pruritic, grouped vesicles and papules, often distributed symmetrically on extensor surfaces such as the elbows, knees, buttocks, and back. It is closely associated with celiac disease and is triggered by gluten ingestion in genetically predisposed individuals with human leukocyte antigen DQ2 and DQ8 haplotypes. Dermatitis herpetiformis is caused by deposition of IgA antibodies that target tissue transglutaminase 3 at the dermal papillae, leading to inflammation and blister formation.[8] Treatment typically involves a gluten-free diet and medications such as dapsone to alleviate symptoms and prevent recurrence.

Subcorneal pustular dermatosis, also known as Sneddon-Wilkinson disease, is a rare chronic relapsing pustular dermatosis characterized by sterile superficial pustules arranged in annular or circinate patterns on erythematous plaques. It predominantly affects middle-aged women and often is associated with underlying

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conditions such as IgA gammopathy or monoclonal gammopathy of undetermined significance. The pathogenesis remains unclear, but immune dysregulation is thought to play a role. Some authors still question whether subcorneal pustular dermatosis is a distinct entity from pustular psoriasis.[4,5,12] Dapsone is the preferred first-line treatment, with adjunct therapies including topical or systemic corticosteroids, other immunosuppressive agents, tumor necrosis factor inhibitors, and UV light therapy.[9]

Definitive management of IH is achieved through early delivery; however, systemic corticosteroids often are used in varying doses to control the disease and to extend the pregnancy period closer to term or until delivery is considered viable. Additional therapies that can be considered include infliximab, cyclosporine, and topical corticosteroids, in conjunction with fluid and electrolyte maintenance.[2,4,10] If symptoms persist despite supportive care and pharmacologic intervention, induction of labor or termination of pregnancy may be indicated. In nonbreastfeeding postpartum mothers with persistent disease, therapies commonly used in generalized pustular psoriasis may be given.[11]

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