Phemphigoid gestationis

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DESCRIPTION

A 32-year-old nulliparous woman presented at 33 weeks' gestation with intense pruritus initiated 1 month earlier. She was under treatment for scabies, with no improvement. She was healthy and her pregnancy had had no intercurrences so far. During the physical examination at the emergency room, extensive erythematous plaques with vesicles and bullae could be seen on her abdomen, trunk, neck and all four extremeties (figures 1 and 2).

Due to the severity of her symptoms, she was admitted to the obstetrics ward in order to undergo intravenous (prednisone 20 mg id) and topical corticotherapy, as well as an oral antihistamine (hydroxyzine 25 mg id). The full blood count and biochemical work-up revealed mild anaemia with no changes in liver function markers, through which intrahepatic cholestasis of pregnancy was excluded as a diagnosis.

A dermatology consult was requested, in which context a biopsy was performed. The histology report was inconclusive in differentiating between phemphigoid gestationis and polymorphic eruption of pregnancy; because of this, an ELISA test was done, which was positive for BP180 antibodies (>200 UA/mL, reference value <20 UA/mL) and direct immunofluorescence confirming the diagnosis of phemphigoid gestationis.

The patient was discharged 1 week later under oral therapy including, once again, a corticosteroid and an antihistamine. At 38 weeks, she delivered a male newborn of 2980 g with no skin lesions. On her third day post partum, however, she had a new flare-up, having to undergo 3 months of therapy with subcutaneous goserelin. In the 4 years since, she has not showed any signs of recurrence.

Phemphigoid gestationis is a rare (1:5000) autoimmune dermatosis of pregnancy, although it can persist through puerperium, as was the case with this patient, and/or be exarcebated with menses and oral contraceptive intake.[1] The blisters may appear on both the mother and the newborn (10% of all cases), but here only the mother was affected.

Pruritus is the main complaint in phemphigoid gestationis.[2] One of its most important differential diagnoses is polymorphic urticarial papules and plaques of pregnancy: in the present case, both histology and direct immunofluorescence were needed to confirm the suspicion. Corticosteroids are the first-line systemic therapy for the disease.

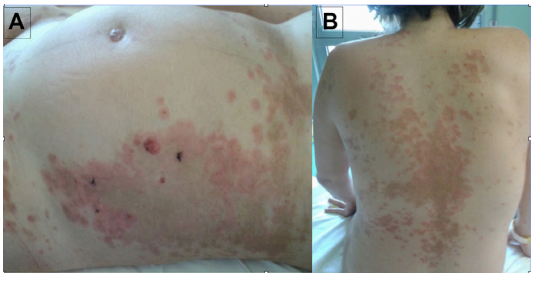

Figure 1 Macroscopic skin lesions of 33-week pregnant woman: initially, lesions are like erythematous papules or targetoid lesions (A, abdomen; B, back).

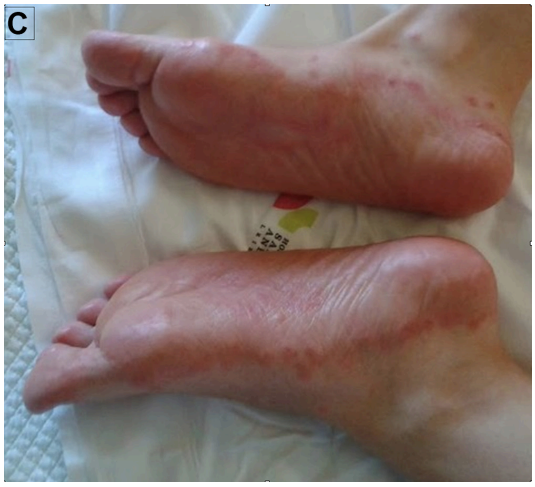

Figure 2 Macroscopic skin lesions of 33-week pregnant woman: over time, small vesicles or bullae appear on normal skin or on top of urticarial plaques (C, feet).

Learning points

- This pregnancy-related blistering dermatosis is associated with poorer obstetric outcomes, like fetal growth restriction and prematurity.
- Immunofluorescence plays an essential role in diagnosis, showing linear deposits of complement component C3 along the basement membrane.
- Women should be informed of the disease's risk of recurrence in subsequent pregnancies, which is 5%–8%.

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