Pustular psoriasis of pregnancy with paradoxical facial involvement: an uncommon presentation-what is your diagnosis?

A 30-year-old primigravida with a gestation period of 34 weeks presented to the dermatology outpatient department with multiple, painful and mildly pruritic, pus-filled lesions of two weeks duration associated with mild fever. The lesions initially started over the chest area as an erythematous rash, followed by development of pustules and then spread rapidly to involve the entire trunk, and upper and lower limbs within five days. There was no history of prior psoriatic lesions. She was not taking any medication except for iron and folic acid. On clinical examination, her vitals were stable. Cutaneous examination revealed erythematous plaques with superimposed pustules over the entire abdomen, trunk, and upper and lower extremities (Figure 1). Her face was also involved, with multiple pustules over the forehead, cheeks and chin. Erythematous plaques with tiny pustules along the margins were seen in both the periorbital areas (Figure 2). Old lesions showed desquamation and scaling. Palms, soles and oral cavity were normal. Obstetric examination showed fundal height consistent with 32 weeks gestation with fetus in cephalic presentation and normal liquor. The patient was admitted under the Dermatology department, with obstetric team taking care of fetal surveillance. Complete blood count showed neutrophilic leukocytosis (10,800/mm^3), low-albumin (2.9 g/dL) and raised erythrocyte sedimentation rate (60 mm/hr). Other laboratory investigations, including corrected calcium, blood sugar, renal and liver function tests, and serum

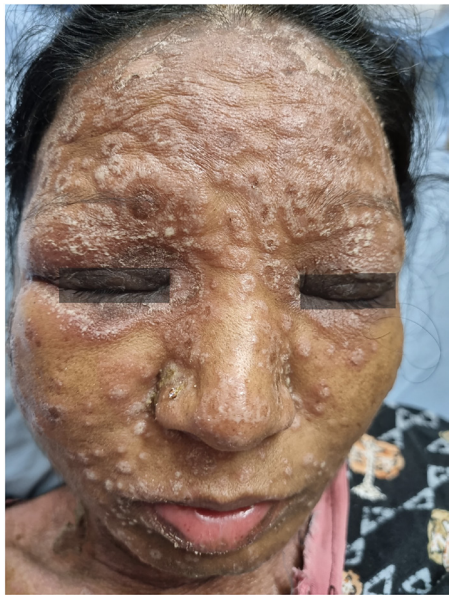

Figure 1. Multiple erythematous plaques with superimposed pustules over entire abdomen, old pustules over upper abdomen showing desquamation

Received: 29 August, 2024 **Accepted:** 23 December, 2024 **Publication Date:** 12 March, 2025

Address for Correspondence: *Mishu Mangla*
e.mail*: mishusingla83@gmail.com* ***ORCID:*** *orcid.org/0000-0003-0708-1037*

DOI: *10.4274/jtgga.galenos.2024.2024-8-2*

Cite this article as: *Konda D, Mangla M, Saisumanth T, Palo S, Kumar N. Pustular psoriasis of pregnancy with paradoxical facial involvement: an uncommon presentation-what is your diagnosis? J Turk Ger Gynecol Assoc. 2025; 26: 62-4*

electrolytes were within normal limits. Swabs from the pustules were sterile and showed no growth. Skin biopsy from an involved area showed intracorneal and sub-corneal pustule formation with marked neutrophilic infiltrate. The epidermis also showed hypogranulosis, focal parakeratosis, and irregular acanthosis with mild supra-papillary thinning at places. The superficial dermis showed few dilated capillaries, and perivascular inflammation comprised of neutrophils and a few lymphocytes confirming the diagnosis of pustular psoriasis of pregnancy (PPP) (Figure 3).

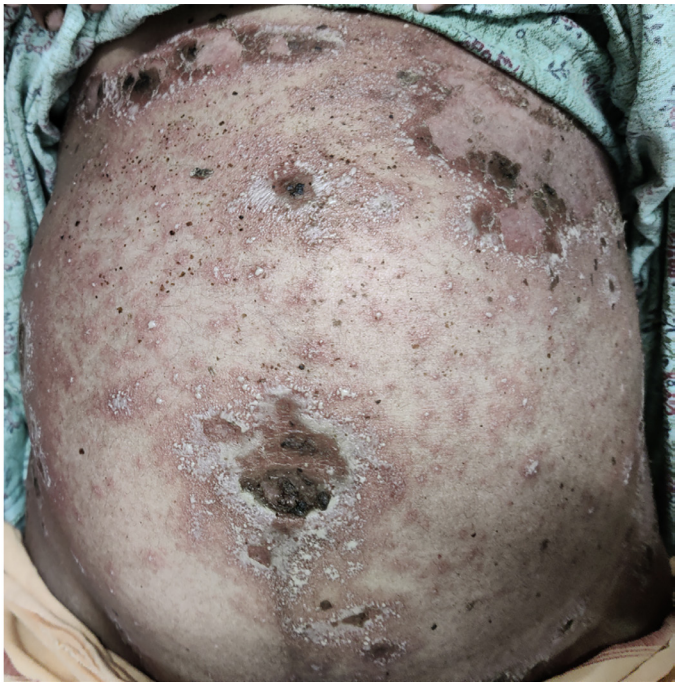

Figure 2. Multiple pustules arranged in annular pattern over underlying erythema over forehead, cheeks, chin and peri-orbital areas

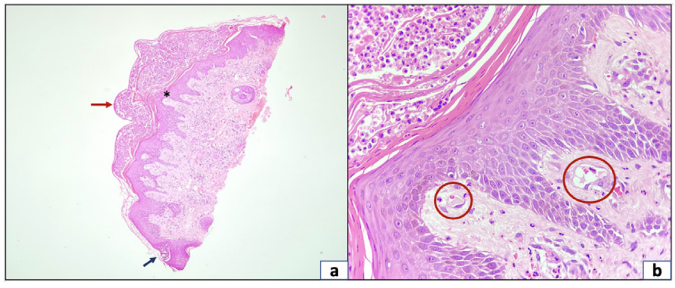

Figure 3. (a) Low power view showing intra-corneal (red-arrow) and sub-corneal (black-arrow) pustule formation, irregular acanthosis, elongated rete-pegs and focal thinning of supra-papillary epidermal plates (asterisk); (b) Higher magnification showing neutrophilic content of the intra-corneal pustule, underlying parakeratosis, hypogranulosis, mild spongiosis, and dilated capillaries in the papillary dermis (circled)

The patient was started on oral prednisolone 60 mg along with protein supplementation. Initially lesions subsided but after the third day new eruptions of pustules developed and showed no improvement even after one week. So, she was started on oral cyclosporine 150 mg (3 mg/kg/day) per day with regular monitoring of blood pressure, renal parameters, and serum electrolytes. Due to the risk of utero-placental insufficiency in PPP, she was kept on strict antepartum surveillance with weekly Doppler and non-stress test. Lesions started desquamating on day two and there was complete resolution of the lesions by the tenth day. However, on tapering the cyclosporine dose, she developed new pustules. So, she was maintained on 150 mg of cyclosporine until 38 weeks of pregnancy. Labor was induced at 38 weeks and she delivered a healthy female child with birth weight 2260 grams. After delivery, cyclosporine was tapered and stopped over a period of five weeks. The patient was reviewed one month after stopping cyclosporine and there were no new pustules.

Answer

PPP, previously known as impetigo herpetiformis (IH), is a rare, severe pregnancy dermatosis that carries a bad prognosis for both the mother and the baby if left untreated. In 1872, Von Hebra et al. (1) first coined the incorrect name IH after reporting five pregnant women with inflammatory clustered pustular lesions with five fetal and four maternal deaths. Later, in 1910, Leo Ritter von Zumbusch (1874-1940), an Austrian dermatologist and professor from Vienna, first described the term, generalized pustular psoriasis (GPP) (2). Currently, the term IH is unfavored and is being replaced by the term PPP by many authors as it is most likely to be a variation of GPP that flares up in response to a variety of triggers present in pregnancy. These include metabolic disturbances, systemic steroid withdrawal, and pregnancy itself. PPP is also considered to be a dermatosis of pregnancy owing to the importance of early recognition and treatment (3).

The exact pathophysiology of PPP is not currently known. Pathogenic mutations in the interleukin 36 receptor antagonist (IL36RN) gene, personal or family history of psoriasis, low serum vitamin D, increasing levels of progesterone during the last trimester of pregnancy, hypocalcaemia, and disruption in elastase activity are some of the factors known to be associated with PPP (3). PPP usually occurs in the third trimester of pregnancy, as in the described case, owing to increased progesterone levels. However, it has also been reported to occur during first trimester and postpartum. PPP is also known to recur in a significant proportion of subsequent pregnancies and that too at an early period (4). Clinically, PPP is characterized by erythematous plaques with pustules along the margins. The lesions initially develop in skin folds and then gradually spread

centrifugally to involve the entire body. The face, palms and soles are typically spared with occasional involvement of oral mucosae. Our patient had atypical involvement of her face. Skin lesions are usually associated with systemic features, including fever, myalgia and malaise (2).

The lesions of PPP are known to resolve spontaneously after parturition. However, it can result in life threatening complications if not treated and monitored properly during pregnancy. Various complications include cardiovascular failure, severe respiratory distress syndrome, hypocalcemia seizures, tetany, hypoalbuminemia, delirium in the mother and placental insufficiency, intrauterine growth restriction, premature rupture of membranes, miscarriage, fetal abnormalities, and stillbirth in the baby (3). Differential diagnoses of PPP include pustular psoriasis, acute generalized exanthematous pustulosis, sub-corneal pustular dermatosis, gestational pemphigoid, and atopic eruption of pregnancy (5).

Aggressive treatment and close monitoring of the mother and fetus are vital in the management of women with PPP. Oral corticosteroid, especially prednisolone, is considered to be the first choice of drug in mild cases of PPP. The dose of prednisolone can vary from 30 mg to 80 mg depending upon the response. However; in cases resistant to steroid and in severe cases, oral cyclosporine at a dose of 3-5 mg/kg/day has become the accepted therapy. Moreover, the efficacy of cyclosporine is enhanced when combined with oral prednisolone. Biological agents like infliximab and secukinumab, and narrow band ultraviolet-B have also been used to treat PPP (2,4,6).

Thus, we present an atypical case of PPP with a rare distinguishing feature of severe facial involvement in the form of pustules and erythematous plaques with pustules at the margin in the periorbital areas.

Deepthi Konda[1], Mishu Mangla[2], Thota Saisumanth[1], Seetu Palo[3], Naina Kumar[2]

[1]Department of Dermatology, All India Institute of Medical Sciences, Bibinagar, Telangana, India

[2]Department of Obstetrics and Gynaecology, All India Institute of Medical Sciences, Bibinagar, Hyderabad, India

[3]Department of Pathology and Laboratory Medicine, All India Institute of Medical Sciences, Bibinagar, Telangana, India

Ethics

Informed Consent: *Informed consent was obtained from the patient for the publication of case details and associated images.*

References

1. Von Hebra F. On some affections of the skin occurring in pregnant and puerperal women. Wien Med Wochenschr. 1872; 22: 1197-202.
2. Ennouri M, Bahloul E, Sellami K, Marrakchi S, Fakhfakh F, Turki H, et al. Pustular psoriasis of pregnancy: clinical and genetic characteristics in a series of eight patients and review of the literature. Dermatol Ther. 2022; 35: 8-10.
3. Namazi N, Dadkhahfar S. Impetigo herpetiformis: review of pathogenesis, complication, and treatment. Dermatol Res Pract. 2018; 2018: 5801280.
4. Joshi KS, Mohammad S, Acharya N, Joshi S. Impetigo herpetiformis complicating pregnancy: a case report on a rare gestational dermatosis with constitutional symptoms. Cureus. 2023; 15: e47898.
5. Trivedi MK, Vaughn AR, Murase JE. Pustular psoriasis of pregnancy: current perspectives. Int J Womens Health. 2018; 10: 109-15.
6. Zhang J, Xia P, Wan L, Chen L, Zhou X, Chen J. Generalized pustular psoriasis of pregnancy successfully treated with secukinumab. Indian J Dermatol Venereol Leprol. 2023; 89: 886-8.