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Case Report

Pustular autoimmune progesterone dermatitis in pregnancy: a rare presentation

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ABSTRACT

Autoimmune progesterone dermatitis (AIPD) is a rare, hormonally mediated dermatosis characterized by recurrent premenstrual skin eruptions caused by hypersensitivity to endogenous or exogenous progesterone. Although symptoms often subside during pregnancy due to hormonal adaptation, re-exposure to progesterone can trigger disease flares. A 30-year-old woman presented with complains of generalized erythematous and itchy eruption with few pustular lesions on the face for 5 days. Upon enquiring she revealed that she is 2 months pregnant and started on oral micronized progesterone prescribed for obstetric support. Laboratory investigations were normal, and histopathology showed a perivascular lymphocytic infiltrate with dermal edema, suggestive of hypersensitivity. Based on the temporal association between progesterone exposure and lesion recurrence, a diagnosis of autoimmune progesterone dermatitis was established. The withdrawal of progesterone, along with treatment using antihistamines and a short course of corticosteroids, resulted in complete resolution. The patient remained symptom-free throughout pregnancy and postpartum, with no recurrences during 18 months of follow-up. This case underscores the diagnostic significance of exogenous progesterone induced autoimmune progesterone dermatitis. Awareness of this rare condition is essential, particularly during pregnancy, for prevention and effective management.

Keywords: Autoimmune progesterone dermatitis, Exogenous progesterone, Hypersensitivity

INTRODUCTION

Autoimmune progesterone dermatitis (AIPD) is an uncommon hormone-related dermatosis presenting with recurrent skin and mucosal eruptions that typically appear during the luteal phase of the menstrual cycle and resolve within one to two days after menstruation.[1] The exact mechanism remains uncertain; however, it is thought to represent a hypersensitivity reaction to the elevated endogenous progesterone levels seen post-ovulation.[2] The true incidence of AIPD is unknown, with fewer than 200 cases have been reported in the literature.[3] Here, we present a case of AIPD which presented in first trimester of pregnancy with the use of exogenous progesterone, and provide an updated review of its clinical presentation, underlying mechanisms, diagnostic approach, and therapeutic options.

CASE REPORT

A 32 years old female presented with widespread erythematous, itchy papules and plaques over the trunk, extremities, and back, accompanied by a few tiny pustular lesions over the face for 5 days. (Figure 1A and B) There were no mucosal ulcers or systemic symptoms. There is no previous history of such lesions. Upon enquiring she revealed that she is 2 months pregnant and started on oral micronized progesterone 7 days back by her obstetrician

for obstetric support. Within 48 hours of starting the medication, she developed above mentioned symptoms. Laboratory investigations including complete blood count, liver and renal function tests, thyroid profile, and autoimmune screening (ANA, anti-dsDNA, anti-Ro/SSA) were within the normal limits. A skin biopsy from a trunk lesion revealed perivascular lymphocytic infiltrate with mild dermal edema, consistent with urticarial hypersensitivity. Based on clinical presentation following progesterone administration, lab investigations, final diagnosis of AIPD was made.

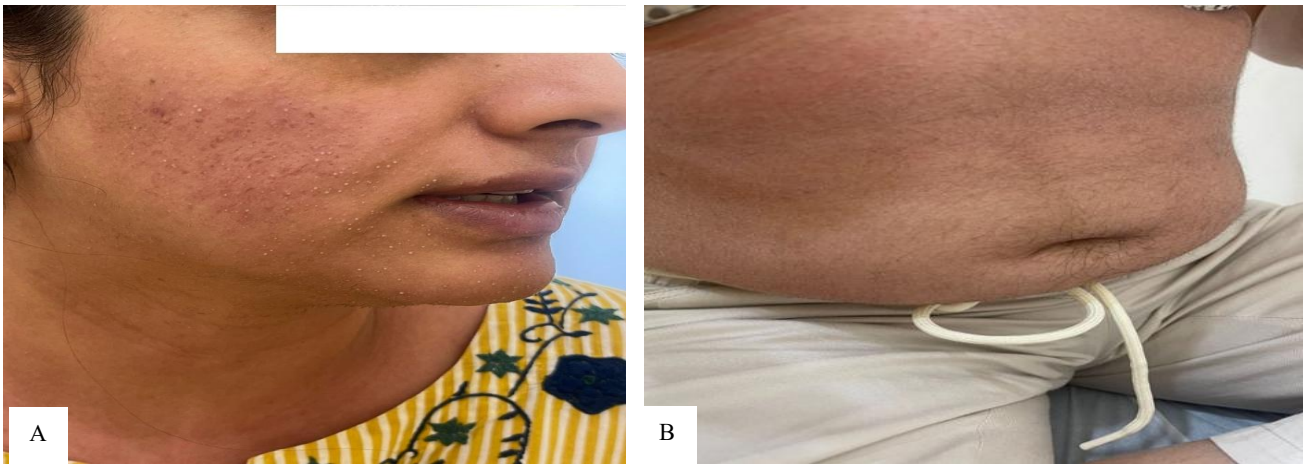

Figure 1 (A and B): Widespread erythematous, itchy papules and plaques over the trunk, extremities, and back.

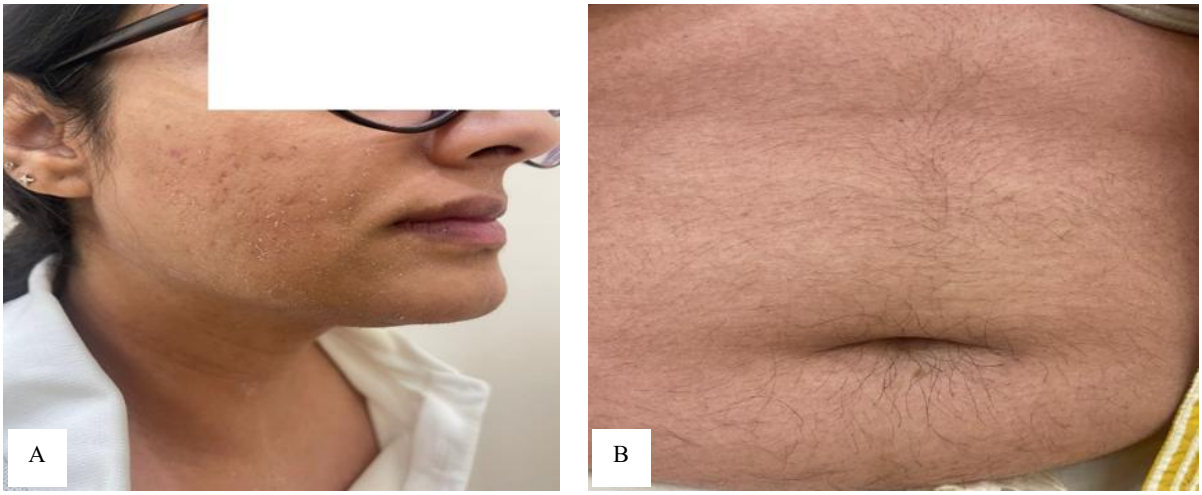

Figure 2 (A and B): Post inflammatory desquamation seen over cheeks and resolving erythema over abdomen.

The progesterone supplementation was immediately discontinued, and she was managed conservatively with oral antihistamines (chlorpheneramine 4 mg bd per day) and a short course of low-dose systemic corticosteroids (prednisone 20 mg/day, tapered over one week). The cutaneous lesions resolved completely within five days. Post inflammatory desquamation seen over cheeks (Figure 2A) and resolving erythema over abdomen (Figure 2B).

The remainder of her pregnancy was uneventful, and she delivered a healthy term infant without obstetric or neonatal complications. During the postpartum period, the patient remained asymptomatic while breastfeeding. At one-year follow-up, she was free of any cutaneous lesions with no recurrence of lesions.

DISCUSSION

AIPD also termed progesterone hypersensitivity, is a rare hormone-mediated dermatosis characterized by recurrent cutaneous and/or mucosal eruptions temporally related to progesterone exposure.[4] The earliest report of menstruation-associated cyclic urticaria was documented by Géber in 1921.[5] Although historically linked to endogenous progesterone fluctuations during the luteal phase, but increasing evidence suggests that exogenous progesterone plays a significant role in triggering or unmasking the disease.[6]

The pathogenesis of AIPD remains incompletely elucidated. Current literature supports both type I (IgE-mediated) and type IV (cell-mediated) hypersensitivity

reactions directed against progesterone or progesterone–protein complexes. Sensitization following exposure to exogenous progesterone with subsequent cross-reactivity to endogenous progesterone has been proposed as a key mechanism.[7] The rapid onset of symptoms within 48 hours of initiating oral micronized progesterone in our patient, in the absence of prior cyclic symptoms, strongly supports an exogenous progesterone-induced hypersensitivity reaction as reported in previous studies.[6]

Clinically, AIPD is highly polymorphic, with reported presentations including urticaria, eczematous dermatitis, erythema multiforme–like lesions, vesiculobullous eruptions, pustules, and anaphylaxis.[6] However, the facial pustulation observed in our patient is a rare presentation.

AIPD presenting during pregnancy is particularly rare. Pregnancy may exert variable effects on disease activity, with reports of both symptom remission due to progressive progesterone desensitization and disease exacerbation or new onset, especially following progesterone supplementation.[8,9]

Diagnosis is primarily clinical, based on the temporal association with progesterone exposure and symptom resolution after cessation. Diagnostic criteria proposed by Warin et al include lesions on the skin accompanied with onset of menstrual cycle, a positive intradermal test, and improvement of symptoms after treatment with progesterone inhibition therapies.[10] The intradermal test can support the diagnosis of AIPD based on the symptoms; on the contrary, in asymptomatic women, a positive intradermal test can result in false negative; therefore, this test might not be the most efficient diagnostic criteria for diagnosis of AIPD. Histopathological findings are nonspecific and typically reflect a hypersensitivity reaction.

Therapeutic options for AIPD focus on symptomatic relief and hormonal modulation. Antihistamines and low dose corticosteroids—topical or systemic—are commonly used for symptomatic control. Long-term management aims to inhibit ovulation and reduce endogenous progesterone levels. Combined oral contraceptives are typically first-line therapy, while gonadotropin-releasing hormone (GnRH) agonists, danazol, or tamoxifen may be used in resistant cases. Surgical options such as bilateral oophorectomy have been reported for refractory or severe disease unresponsive to medical therapy.[11]

CONCLUSION

This case highlights the occurrence of autoimmune progesterone dermatitis during pregnancy with exogenous progesterone exposure without any previous history of lesions during menstrual cycles. Early recognition and avoidance of progesterone-containing medications are crucial for preventing exacerbations in such patients.

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