

Autoimmune Progesterone Dermatitis: A Case Report

Tahani Magliah (1)

Salwa Bardisi (2)

(1) Dermatology specialist

(2) Family medicine consultant

Corresponding author:

Dr. Tahani Magliah

Makkah Al-Mukarramah City,

Saudi Arabia

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Abstract

In this case report, we present a 29-year old female, who complained of a recurrent cyclical itchy skin rash over the dorsum of her hands for the last four years. She used to develop the rash during the initial weeks of each pregnancy, which then subsided spontaneously. The symptoms markedly decreased when she used contraceptive pills. On examination, there were multiple scaly erythematous plaques over the dorsum of the hands with signs of lichenification. Intradermal progesterone test showed an itchy erythematous papule over a wheal, at the site of injection, sized about one cm, which appeared after 48 hours. Therefore, she was diagnosed as a case of "autoimmune progesterone dermatitis". Daily oral contraceptive pills (levonorgestrel/ethinyl estradiol, 0.1 mg to 20 µg) were prescribed. There was a marked reduction in the number of lesions during her next menstrual periods.

Key words:

Autoimmune progesterone dermatitis, progestogens hypersensitivity, Intradermal progesterone test, contraceptive pills.

Introduction

Autoimmune progesterone dermatitis (APD) is one of the rare dermatoses. It is characterized by recurrent skin eruptions that appear during the luteal phase of the menstrual cycle (1). The nature of skin eruption varies and is diverse, and so, its diagnosis is challenging. But with a history of recurrent cyclical skin eruptions and symptomatic improvement after inhibition of progesterone, with an intradermal positive result of progesterone test can help to confirm the diagnosis (2).

In this case report, we present a rare case of a mother with APD dermatitis, who is probably the first documented case of APD to be reported in Saudi Arabia.

Case Report

A 29-year old female, not known to have any medical illness, came complaining of recurrent skin rash for the last four years. The rash is characterized by its cyclical pattern, itchy eczematous-like over the dorsum of the hands.

The patient denied history of any food or drug allergy, recent medication use, or history of animal contact. She is married and a mother of two children. During her pregnancies, she used to develop the same rash in a few initial weeks of each pregnancy, which then subsided spontaneously. Moreover, when she used contraceptive pills between the two pregnancies, the rash and the symptoms markedly decreased.

She visited several dermatology clinics but her condition was diagnosed as atopic dermatitis, contact dermatitis or urticaria. She was advised to avoid stress and to use topical steroids and antihistamine, but minimal improvement occurred.

When the patient was examined, there were multiple scaly erythematous plaques over the dorsum of the hands with signs of lichenification, indicating prolonged rubbing due to itching (Figures 1,2). There were no other skin areas or mucosal involvement.

Intradermal progesterone test showed an erythematous papule over a wheal, sized about one cm at the site of injection, itchy, which appeared after 48 hours (Figure 3).

Therefore, our patient was diagnosed as a case of "autoimmune progesterone dermatitis" based upon her history, physical examination and the positive results of the intradermal progesterone skin test.

Since the patient wanted to postpone pregnancy, daily oral contraceptive pills (levonorgestrel/ethinyl estradiol, 0.1 mg to 20 µg) were prescribed to her and she was asked to come to our clinic every month for follow up.

There was a marked reduction in the number of lesions during her next menstrual period. After three menstrual cycles, no lesions appeared peri-menstrually. Our patient continued using levonorgestrel/ethinyl estradiol for a total of six months, after which her symptoms were successfully controlled. Afterwards, her lesions did not return despite the discontinuation of contraceptive pills.

Figures (1,2): Erythematous scaly plaques over the dorsum of both hands with signs of lichenification



Figure 1



Figure 2



Figure 3: Erythematous papule surrounded by wheal at the site of intradermal progesterone injection, which appeared after 48 hours

Discussion

Our patient presented with a recurrent cyclical itchy eczematous-like skin rash over the dorsum of her hands, which was characterized by its cyclical behavior. She noticed that her rash used to flare during the initial weeks of each pregnancy, then partially subside. Moreover, after labor, her complaints markedly decreased when she used contraceptive pills.

Gupta et al. (3) stated that hypersensitivity responses to progestogens occur among women during their reproductive age and can present with a heterogeneous group of skin and/or systemic reactions that are temporally associated with relative peaks of serum progesterone levels. These reactions are usually affected by sources of progesterone, endogenously or exogenously.

The menstrual cycle has been associated with a variety of skin eruptions, including eczema, prurigo, erythema multiforme, stomatitis, bullous-vesiculo pustular eruptions, folliculitis, angioedema, and urticarial plaques (4). The first documented case of APD was in 1921, in which a patient's premenstrual serum caused acute urticarial lesions (5).

The association between pregnancy and onset of symptoms in cases of APD has been reported by Nguyen and Razzaque Ahmed (6), who noted that out of 89 cases of APD, 13 were related to a pregnancy, while 7 occurred during their postpartum period. Yavuz et al. (7) stated that pregnancy can interfere with symptoms of APD. This can be explained by that pregnancy is associated with an increase in maternal progesterone levels by 10 to 5,000 times relative to non-pregnancy levels (8).

Our patient went to several dermatology clinics, for which her condition was misdiagnosed as atopic dermatitis, contact dermatitis or urticaria. However, her diagnosis could be reached based upon her history, physical examination and the positive results of the intradermal progesterone skin test.

It has been observed that among some women, there are several skin diseases that may be associated with their menstrual cycle which has been associated with the spectrum of skin diseases, e.g., eczema, erythema multiforme, folliculitis, angioedema, and urticaria. These lesions may appear as typical urticaria or erythema multiforme in various stages of healing. These lesions may be present on the lips, palms of hands, trunk, or feet (8).

However, the diagnostic criteria for APD were proposed by Warin (9), which include: skin lesions associated with the menstrual cycle (premenstrual flare); a positive progesterone intradermal test; and the improvement of symptoms after inhibiting progesterone secretion by suppressing ovulation.

Our patient stated that before being diagnosed, she received antihistamines, but with no marked improvement. However, symptoms were successfully controlled after receiving oral contraceptive pills for a total of six months.

Yavuz et al. (7) stated that, although antihistamines are considered first line therapy, APD is usually resistant to antihistamines. Several treatments that suppress ovulation can be used to control APD. Oral contraceptive pills are the most commonly used therapy. Even a short course of oral contraceptive pills can provide long-lasting improvement of the condition, probably due to desensitization to progesterone (10-11). Danazol has been reported to be effective in some cases. Gonadotropin-releasing hormone agonists were also used since their administration inhibits ovulation and decreases sex hormone production (11). However, they should not be used for more than six months due to their negative effects on bone metabolism and the cardiovascular system (12). Tamoxifen is another therapeutic agent that is used to suppress ovulation and improve symptoms (8). In refractory cases, and as a last resort, bilateral oophorectomy was done (13).

In conclusion, autoimmune progesterone dermatitis is a rare cyclical dermatosis. Its differential diagnosis includes atopic dermatitis, contact dermatitis and urticaria. Its diagnosis is based upon history, physical examination and positive results of intradermal progesterone skin test. Several treatments that suppress ovulation can be used to control APD, but oral contraceptive pills are the most commonly used therapy.

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