

Correspondence

A case of pustular psoriasis possibly precipitated by periodic oestrogen/gestagen therapy for Turner syndrome

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Patients with Turner syndrome (TS) have an X chromosome abnormality, having only one X chromosome, partial deletion of the X chromosome or mosaicism with an abnormal X chromosome. To compensate for the ovarian dysfunction and to protect bone density, oestrogen/gestagen periodic therapy (Kaufmann therapy; KT) is essential for patients with TS.

A 33-year-old woman presented with a recalcitrant rash. She had received growth hormone therapy for short stature until 20 years of age. She had been diagnosed with TS at the age 30 years, and had been started on KT (0.626 mg of conjugated oestrogen on day 1–25 and 5 mg of medroxyprogesterone acetate on day 14–25; followed by menstruation interval). Her karyotype was 46, Xi(X)inv(1)(p13q21). Family history for plaque psoriasis was negative. Two years after initiation of KT, the patient developed persistent multiple erythematous circular lesions with desquamation and pustules at the periphery, along with fever (37.6 °C), general malaise, knee and ankle pain, and marked lower leg oedema (Fig. 1a–c).

Skin biopsy showed mild acanthosis and elongation of rete-ridges with Kogoj spongiform pustule in the epidermis (Fig. 1d). Superficial perivascular cell infiltration, primarily neutrophilic infiltration, was noted in the dermis.

Laboratory examination showed an increased white blood cell count (9760/mm³; normal range 3170–8400/mm³), with high normal level of neutrophils (71%; 39.7–71.2%) and high C-reactive protein (1.6 mg/dL; 0–0.3 mg/dL).

The patient was diagnosed with generalized pustular psoriasis (GPP). KT was discontinued. Oral ciclosporin 125 mg (3.5 mg/kg) and maxacalcitol/betamethasone butyrate propionate (as combination formula) were effective at relieving the symptoms of skin eruptions and joint pain within 7 days. The ciclosporin dose was successfully tapered off subsequently (Fig. 1e). Meanwhile, the Kaufmann hormone therapy was resumed two times, and each time, the pruritic pustular eruptions relapsed, and subsided shortly after the therapy was discontinued again (Fig. 1e).

Genetic study was performed following approval of the study protocol by the institutional review board of Nagoya University Graduate School of Medicine and written informed consent from the patient. Sanger sequencing for all exons and flanking introns of *IL36RN*, *AP1S3* and *CARD14* disclosed no pathological mutation.

Patients with TS are susceptible to various autoimmune diseases, and the morbidity rate of patients with TS and psoriasis is 5.4 times higher than that in healthy individuals with psoriasis.¹ In addition, the high progesterone state is reportedly associated both with GPP and impetigo herpetiformis.^{2,3} In the present case, KT was restarted two times, and the eruption recurred soon afterwards. Thus, it is reasonable to speculate that KT triggered GPP in this case.

The mutation analysis for *IL36RN*, *AP1S3* and *CARD14* was negative, and this may explain why the GPP was relatively mild. A literature search identified six other cases of GPP associated with Turner syndrome.⁴ All these cases were mild and responsive to treatment, as in our case. Mutation analysis was performed in one case, and was also negative.⁵ Whether the absence of mutations in *IL36RN*, *AP1S3* and *CARD14* is common in patients with TS and GPP warrants further study.

T. Shiratori,¹ T. Takeichi,² M. Kono,² M. Nishida,¹ A. Imanishi,¹ N. Maekawa,¹ N. Kawamura³ and K. Fukai¹

¹Departments of Dermatology; ³Gynecology, Osaka City General Hospital, Osaka, Japan and ²Department of Dermatology, Nagoya University Graduate School of Medicine, Nagoya, Japan
E-mail: hewuiewskau0346@yahoo.co.jp

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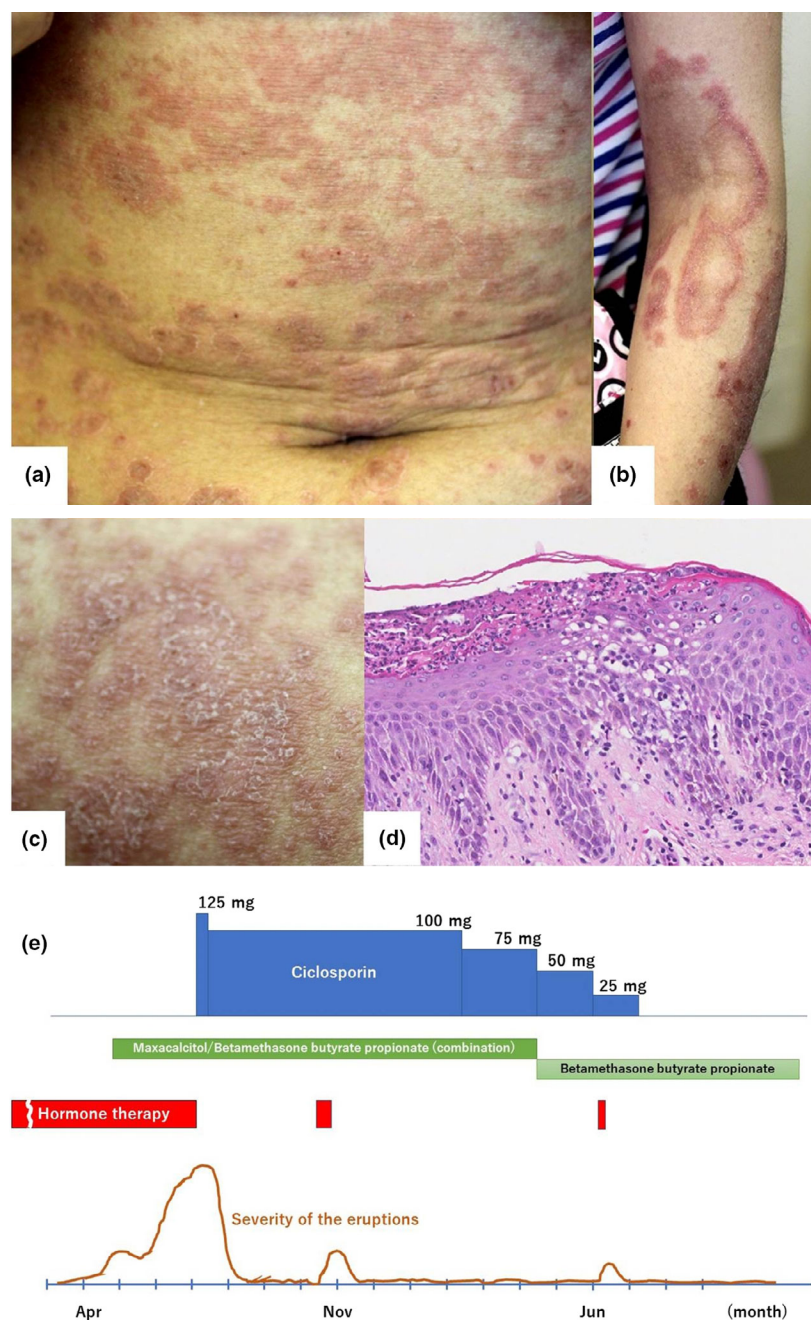


Figure 1 (a) Multiple coalescent infiltrated erythematous lesions with desquamation on the trunk; (b) annular erythema with peripheral pustules on the extremities; and (c) small pustules within the infiltrated erythema on the trunk. (d) Histological examination result revealed accumulation of neutrophils under the stratum corneum and formation of a thick crust, and neutrophils were present within the spongiotic epidermal cells, forming Kogoj spongiform pustules (haematoxylin and eosin, original magnification $\times 100$). (e) Time course of progression of severity of the eruptions and the therapy used.

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