Generalized pustular psoriasis of pregnancy treated with oral cyclosporin

Sir,

Generalized pustular psoriasis of pregnancy is a rare variant of pustular psoriasis, also called as impetigo herpetiformis. The disease in its severe and longstanding form has its impact on fetal outcome by causing placental insufficiency resulting in still birth, neonatal death or fetal abnormalities.[1-3] Though the disease remits after parturition during the postpartum phase, recurrences are common in subsequent pregnancy.[4] Systemic steroids are the drugs of choice for the treatment. Cyclosporin is a selective immunosuppressant that has been effectively used in the treatment of pustular psoriasis and is grouped into category C for pregnancy drug risks. Though there are reports of its use in this condition in a few cases, the experience is limited.[5] Our patient was a case of precious pregnancy not showing adequate response to oral steroids that was treated effectively with cyclosporin.

A 25-year-old primigravida of 12 weeks gestation presented with sudden onset crops of pustular lesions of generalized nature. They were associated with pruritus, high-grade continuous fever and chills. Lesions started as pustules on the abdomen and thighs, which then became generalized within three days. She had associated bilateral knee joint pain without any redness or swelling of joints. Her conception had occurred after three years of marriage and hence was precious. Cutaneous examination revealed multiple erythematous plaques studded with multiple grouped pustules at the margins and coalescing at few places to form "lakes of pus"[Figure 1].

Laboratory investigations revealed hemoglobin of 12.8 gm% with total leukocyte count of 9,000 per cmm. Liver function tests, renal function tests, blood sugars, urine, stool examination, VDRL, enzyme-linked immunosorbent assay for human immunodeficiency virus, serum electrolytes, serum calcium and phosphorus were normal. Pustules were sterile and histopathology showed characteristic features of pustular psoriasis. A diagnosis of pustular psoriasis of pregnancy was made and the patient was started on oral prednisolone 60 mg daily. This was given for two weeks and then gradually tapered over a period of two months. However, there was no significant relief and the patient continued to get showers of new pustules. This was noticed especially once the dose of steroid was tapered below 20 mg/day. Hence at 26 weeks of gestation oral cyclosporin was introduced at the dose of 50 mg bid (2 mg/kg). Obstetrician reference for fetal monitoring was done at regular intervals. With cyclosporin her constitutional symptoms and lesions came under control.

At about 32 weeks of gestation, there was exacerbation of lesions with high-grade fever, which

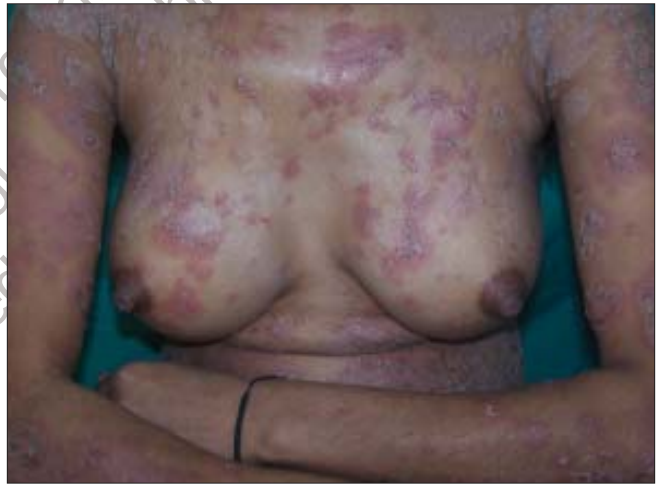

Figure 1: Impetigo herpetiformis

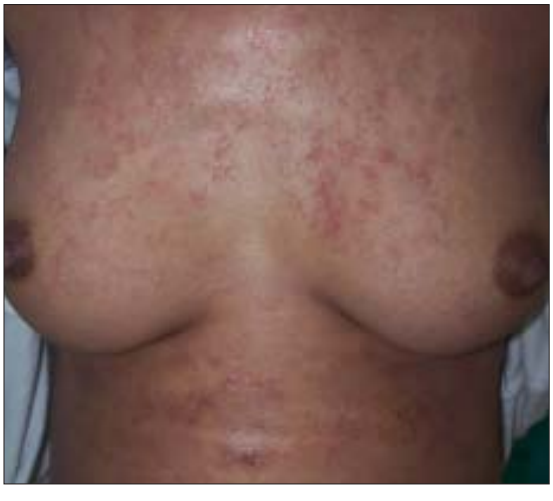

Figure 2: Marked improvement in the lesions after 8 weeks of cyclosporin therapy

was followed next day by preterm delivery of a 1.6 kg male infant. Preterm baby was shifted to the neonatal intensive care unit. Oral cyclosporin was continued. During the postpartum period by the second week most of the skin lesions improved and by the end of a month lesions subsided completely with areas of hyperpigmentation [Figure 2].

Impetigo herpetiformis was first described by von Hebra in 1872 in five pregnant women, four of whom had died.[6] and by 1982 about 200 cases were reported.[4] This rare pustular eruption tends to occur commonly in the third trimester of pregnancy although cases have been reported as early as the first trimester.[7] Most of the affected cases had no previous or family history of psoriasis as in our case. The exact etiology is still not known but the role of high progesterone levels during the last trimester, low levels of calcium and reduced amount of skin-derived antileukoproteinase activity were proposed in the pathogenesis.[8,9]

Recurrence is common in subsequent pregnancies and upon subsequent use of oral contraceptives.[1,4] Fulminant disease in pregnancy is best treated with prednisolone in doses up to 60 mg as was done in our case. However, on tapering the steroids, the condition relapsed prompting us to switch to cyclosporin. With the introduction of cyclosporin the constitutional symptoms were much less and the pustular eruptions were under control.

REFERENCES

1. Oumeish OY, Farraj SE, Bataineh AS. Some aspects of impetigo herpetiformis. Arch Dermatol 1961;83:103-5.
2. Lotem M, Katzenelson V, Rotem A, Hod M, Sandbank M. Impetigo herpetiformis: A variant of pustular psoriasis or a separate entity? J Am Acad Dermatol 1989;20:338-41.
3. Beveridge GW, Harkness RA, Livingstone JR. Impetigo herpetiformis in two successive pregnancies. Br J Dermatol 1966;78:106-12.
4. Ott F, Krakowski A, Tur E, Lipitz R, Weisman Y, Brenner S. Impetigo herpetiformis with lowered serum level of vitamin D and its diminished intestinal absorption. Dermatologica 1982;164:360-5.
5. Meinardi MM, Westerhof W, Bos JD. Generalized pustular psoriasis (von Zumbusch) responding to cyclosporine A. Br J Dermatol 1987;116:269-70.
6. Hebra F Von. One some affections of the skin occurring in pregnant and puerperal women. Wien Med W Schr 1872;48:1197; abstract in Lancet 1872;1:399, Ann J Syph Dermatol 1873;4:156.
7. Gligora M, Kolacio Z. Hormonal treatment of impetigo herpetiformis. Br J Dermatol 1982;107:253.
8. Katsambas A, Stavropoulos PG, Katsiboulas V, Kostakis P, Panayiotopoulos A, Christofidou E, *et al*. Impetigo herpetiformis during the puerperium. Dermatology 1999;198:400-2.
9. Kuijpers AL, Schalkwijk J, Rulo HF, Peperkamp JJ, van de Kerkhof PC, de Jong EM. Extremely low levels of epidermal skin-derived antileucoproteinase/elafin in a patient with impetigo herpetiformis. Br J Dermatol 1997;137:123-9.

M. M. Kura, A. U. Surjushe
Department of Skin and STD, Grant Medical College and JJ Hospital, Mumbai, India.

Address for correspondence: Dr. Mahendra Kura, Department of Skin and STD, 43, J. J. Hospital, Byculla, Mumbai - 400 008, India.
E-mail: mkura@vsnl.net