Impetigo Herpetiformis: A challenging scenario to the treating dermatologist: A case series

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Egyptian Dermatology Online Journal 10 (2): 5, December 2014

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Abstract

Impetigo herpetiformis (IH) is a rare pustular form of psoriasis in pregnancy, associated with constitutional symptoms and complications of secondary infection and sepsis and an increased risk of fetal abnormalities and stillbirths. Here we present a series of four cases of impetigo herpetiformis, primarily treated with systemic corticosteroids. Dapsone and cyclosporin were added in patients with persistence of skin lesions or showing resistance to steroids. Lesions resolved in all with good obstetric outcome, except in one who had an intrauterine death (IUD) at 34 weeks of gestation despite good response of skin lesions to steroids, providing additional evidence that IH is associated with placental insufficiency, stillbirths and neonatal deaths. In this article we emphasize the importance of early diagnosis and treatment to minimize maternal and fetal mortality and morbidity and considering other therapeutic options like dapsone and cyclosporine as alternative treatment options for IH in case of poor response to corticosteroids.

Introduction

Impetigo herpetiformis is a rare, life threatening pustular dermatoses of pregnancy with an obscure etiology. First described by Hebra in 1872 [1], this disorder is characterized by erythematous plaques with grouped sterile pustules at their margins, starting as symmetrical lesions in intertrigenous areas that extend centrifugally, associated with severe constitutional symptoms. It usually starts in the last trimester, but may occur as early as third month of the pregnancy. Mucous membranes and nails can also be affected. Hypocalcemia is commonly associated with IH. The disease resolves promptly postpartum. The main complications are those of placental insufficiency, an increased risk of still births, fetal abnormalities and neonatal deaths. We present a series of four cases of impetigo herpetiformis, all presenting in the second and third trimester of pregnancy.
Case Reports

Case 1: A 22 year old primigravida, presented at 24 weeks of gestation, with history of generalized pustular eruption for two weeks. Dermatological examination revealed erythematous plaques studded with minute grouped pustules coalescing to form bigger sheets of lesions, distributed over the neck, trunk, arms, dorsum of hands, legs and buttocks (Fig 1). Oral cavity examination revealed geographic tongue.

Fig 1: Impetigo herpetiformis in a 22 year old female. There are multiple ill-defined erythematous scaly plaques with dried up pustules on the abdomen and infra-mammary folds.
Case 2:- A 27 years old primigravida, presented with fever and generalized pustular lesions at 26 weeks of gestation for two weeks. On examination, erythematous plaques with an active polycyclic elevated border comprised of micropustules were distributed on the face, neck, trunk and extremities (Fig 2). Mucosae were normal.

Fig 2: Impetigo herpetiformis in a 27 year old female showing multiple erythematous plaques many of which have peripheral rim of desiccated pustules.
**Case 3:** A 29 years old multigravida, a known case of psoriasis for 7 years, presented at 28 weeks of gestation, with multiple plaque type lesions, studded with pustules for three weeks. The lesions initially appeared over the submammary region that rapidly progressed to involve chest, lower abdomen and limbs (*Fig 3*). Her earlier pregnancy was uneventful. On examination, pinpoint pustules over an erythematous background were present. Mucosae were normal.

*Fig 3:* Impetigo herpetiformis in a 29 years old multigravida showing diffuse silvery white scaling which has occurred due to drying and desiccation of pustules under a background of dull erythema.
**Case 4:** A 21 years old primigravida presented at 28 weeks of gestation, with widespread painful erythematous rash with pustules for the last one month. Dermatological examination revealed widespread erythematous-squamous plaques with tiny superficial pustules. (Fig 4). Geographic tongue with fissures was also present.

**Fig.4a:** Impetigo herpetiformis in a 21 year old primigravida showing widespread scaly erythematous plaques with tiny superficial pustules in frontal view
The lesions were associated with an intense itching and burning sensation and severe constitutional symptoms in all cases. Personal history of psoriasis was present in one patient (case 3), but there was no family history of psoriasis. Systemic and obstetrical examinations were normal.

Laboratory findings revealed leucocytosis and raised ESR in all patients. Liver function tests and renal function tests were within normal limits. Serum calcium level was low in one patient (case 1). Gram stained smear of pustules was negative and culture from the pustules was sterile in all the cases. Histopathology of skin was consistent with pustular psoriasis in all cases. Ultrasonography did not reveal any abnormalities except intrauterine growth retardation in one patient (case 3). Based on typical clinical features and histopathological findings, patients were diagnosed to have impetigo herpetiformis.

In cases 1 and 2, systemic steroids at a dose of 60 mg / day, along with other supportive measures were started. The lesions started regressing within two to three weeks. Dapsone 100 mg/day was added and dose of steroids tapered slowly to a minimum of 10mg/day. The treatment was continued throughout the pregnancy and stopped within few days after delivery.
In case 3, systemic steroids at a dose of 60mg/day were started. Her symptoms were adequately controlled in 2 to 3 weeks. Tapering of steroids was started and the eruption remained stable thereafter. She went into spontaneous labour at 34 weeks, delivering a dead baby prematurely. Steroids were further tapered and stopped.

In case 4, systemic steroids at an initial dose of 60 mg/day for 3 weeks failed to control the disease. Cyclosporine at a dose of 4mg/kg body weight was added in two divided doses. The skin lesions started regressing within 1 to 2 weeks. The corticosteroid dose was tapered gradually to 10 mg/day and along with cyclosporine at 100mg/day were continued, with strict monitoring of all relevant parameters, till delivery at thirty-eight weeks and stopped thereafter.

**Discussion**

There is a considerable debate surrounding the classification of IH. Hebra [1] classified it as specific dermatoses of pregnancy. Some consider it to be an acute phase of generalized pustular psoriasis precipitated by endocrine changes of pregnancy in individuals with subclinical disease [2,3] whereas others describe it as an entity distinct from psoriasis [4].

The pathogenesis of this disease remains unclear but is attributed to high levels of progesterone during the last trimester of pregnancy, low levels of calcium and reduced levels of epidermal skin-derived antileukoproteinase activity [5].

The onset in our patients was between 24 to 26 weeks of gestation as reported earlier [6,7]. Geographic tongue was present in two of our patients, consistent with earlier reports [2]. Cases of IH with hypoparathyroidism have been reported where authors speculated that the patients with latent hypoparathyroidism may become hypocalcemic because of increased demand for calcium in the last trimester of pregnancy [8]. Family history was found in none of our patients, comparable to the findings of Ott et al. [6] but has been reported at other places [9].

Oral corticosteroids are the mainstay of treatment in IH [7]. Two of our patients were treated with dapsone after an initial response to oral corticosteroids. Use of dapsone may be considered in subacute and chronic forms of generalized pustular psoriasis, and may be valuable in atypical variants [10]. It was found to be effective, without any adverse effect to both mother and fetus. In one patient (case 3), marked improvement was seen in skin lesions with steroids but went into spontaneous labour that culminated into an IUD. IH occurring during nine successive pregnancies, with eight fetal deaths has been reported [9]. Of late, cyclosporine as well as methotrexate has been used in IH [11,12]. Cyclosporin has been administered in patients with IH and in most of cases, not as a monotherapy but in combination with low dose systemic corticosteroids. In one of our patients (case 4), unresponsive to steroids, cyclosporine was added, that helped to achieve control of the disease, as has been reported earlier also [13]. Despite placental transfer, cyclosporine seems to be rather safe. Children exposed in utero to cyclosporine have been shown to have normal renal function. Use of the lowest active dose and careful monitoring of the fetus is advisable [14].

**Conclusion**

IH is associated with placental insufficiency posing an increased risk to the fetus. The awareness and recognition of the clinical presentation and careful management is important. Alternative therapeutic options in the form of dapsone or cyclosporine may be considered, when corticosteroids are not enough to control the eruption alone or as a steroid-sparing agent. In any case, total regression of lesions must
be expected after delivery.

References


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