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Case Report

Urticarial rash of pregnancy turning into a rare and scary dermatosis of pregnancy: a case report and review of literature

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ABSTRACT

Dermatosis of pregnancy encompasses a group of skin conditions that occurs due to interaction of multiple factors in the body during pregnancy or during post-partum period. We presented a case of 25-year-old G4P1L1A2 with initial symptom of urticarial rash which later progressed to a rare and a scary dermatosis of pregnancy which became a challenge both to the obstetrician and the dermatologist.

Keywords: Rash, Phemhigoid, Dermatosis, Pregnancy

INTRODUCTION

Pregnancy specific dermatosis includes 4 entities- atopic eruption of pregnancy, intra hepatic cholestasis, and Pruritic urticarial papules and plaques of pregnancy (PUPP) and pemphigoid gestationis. Pemphigoid gestations is the rarest form and occurs in the rate 1: 60,000 pregnancies and due to rare occurrence, the diagnosis and management becomes a challenge. Earlier was known as herpes gestationis as the lesions resembled the herpetic lesions. We presented the rarest form of dermatosis: pemphigoid gestationis which initially started an urticarial rash which progressed to the rarest and scariest form.

CASE REPORT

A 25-year-old G4P1A2L1 at 32 weeks was referred from local hospital for intensely pruritic, erythematous eruptions over abdomen and limb in the last 12 days. Initially there was itching which was predominantly intense over abdomen and severe enough to disrupt the daily routine. Following itching patient developed erythematous rash in the periumbilical area. The lesions progressed to involve the trunk, arms, legs and face over a course of 7 to 8 days. The lesions became edematous with a central dusky hue and peripheral rim of vesicles. No history of similar lesions in previous pregnancy, pre-existing skin disease, drug intake and no allergies elicited. Obstetric history was L1 5-year-old male LSCS done in view of failure of induction. 2 spontaneous abortions one at 12 weeks and the other at 8 weeks. Her last menstrual period was 17 December 2020 and she was 32 weeks at presentation. Her general physical examination revealed mild pallor and rest of all the systemic examination were normal. Local examination revealed multiple well defined erythematous round to oval annular edematous plaques (0.5×0.5 cm) with central dusky hue present on the abdomen and upper limb. Patient started methylprednisolone 30 mg but the dose escalated to 40 mg due to diminished response. Biopsy was taken from the perilesional skin of right arm which revealed subepidermal bulla with plasma cells and few eosinophils and the dermis showed perivascular infiltration by lymphocytes. Direct immunofluorescence revealed deposition of C3 complement and IgG at the basement membrane. Patient was under continuous surveillance by the obstetrician to detect fetal growth restriction (FGR) as well as the dermatologist who was managing the pruritus. At 36.3

weeks patient presented in the emergency with pain and was advised LSCS in view of scar tenderness. Pre-op 100 mg prednisolone IV rescue dose was given. As the previous scar was midline hence the incision was given vertical. The previous angry and inflamed lesions had healed well (Figure 1) hence no difficulty was encountered during LSCS. Patient delivered a male baby of 2.5 kg and the newborn had no lesions on the body on examination. Post-op period was uneventful and the steroids were tapered in the graded manner on weekly basis depending upon the patient response and remission. Stitch removal was done on day 12 and the wound had healed well. There was no post- partum relapse and the steroids were stopped 4 weeks post-partum. The scar marks have faded but still present and perhaps take much longer time to vanish (Figure 2).

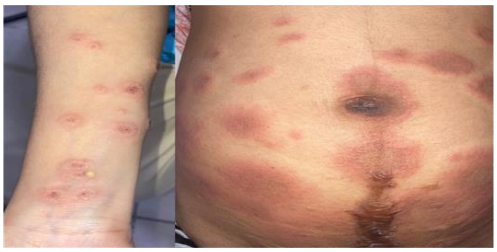

Figure 1: Initial presentation before treatment.

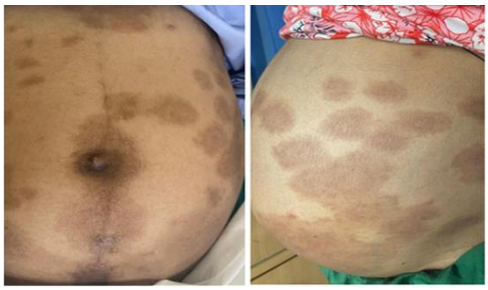

Figure 2: After treatment with steroids.

DISCUSSION

Phemphigoid gestationis is the rarest autoimmune bullous dermatosis of pregnancy. It shares features with bullous dermatosis and generally found in second and third trimester.[2] The recurrence is more severe and can present at an earlier gestational age.

The cause is an aberrant expression of MHC class antigen which causes exposure of BP180 antigen to maternal immune response and results in inflammation and subepidermal blisters.[3] Fluctuation in sex hormones have also been implicated as evident by aggravation of this condition in pregnancy, menstruation and use of oral contraceptive pills. The lesions characteristically start in periumbilical area and involves the trunk, face and the limbs.

The mucosa is generally spared but the mucosa was involved on our case. The lesions start as urticarial rash and later convert into plaques followed by vesicles. Fetal risk encountered are lesions on the neonate (10%), FGR, and prematurity. The maternal risk involved are Grave disease and cosmetic disfugerement.[4] Biopsy is not definitive hence requires direct immunofluorescence as the gold standard which shows complement C3 deposition on basement membrane.[5] It shares very much similarity to PUPP but due to fetal risks involved it needs prompt recognition.

Mild cases are managed by topical steroids and the severe cases response to oral therapy. The safety data of other drugs like azathioprine and immunoglobulins in pregnancy is questionable. Methylprednisolone is safe in pregnancy due to its non\flourinated composition.[6]

The differential diagnosis of dermatosis of pregnancy should be kept in mind while managing and diagnosing this condition.

Table 1: Differential diagnosis of dermatosis of pregnancy.[7]

Lesion	Phemphigoid gestationis	Atopic eruption	PUPPP	Intrahepatic cholestasis
Incidence	Rarest	Most common	-	Common
Parity	Most common	Multi/primi	Primi	Primi/multi
Location	Peri umbilical	Trunk	Peri umbilical sparing	-
Gross morphology	Plaques, vesicles, bullae	Papules, nodules	Papules, plaques	Scar marks
Lab diagnosis	DIF +ve	Clinical HPE	DIF -ve	LFT/bile acid
Maternal/fetal risk	M/F:+/+	M/F:-/-	M/F:-/-	M/F:+/+
Recurrence	+	+	-	+

CONCLUSION

Phemphigoid gestationis is rarest dermatosis and recquires a keen eye to diagnose it due to fetal risks involved. The differential diagnosis should be kept in mind and the multidisciplinary approach could result in favourable outcomes.

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