CASE REPORT

Pregnancy-Associated Sweet's Syndrome: A Rare Clinical Entity

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About the Author

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Introduction

Sweet's syndrome also known as acute febrile neutrophilic dermatoses is characterized by fever, painful cutaneous eruption, neutrophilia and a classical histopathological picture of diffuse neutrophilic infiltrate in the upper dermis. It may be idiopathic or may be associated with infection, drugs, malignancy and pregnancy. Sweet's syndrome can be rarely associated with pregnancy [1]. Here in we report

a case of Sweet's syndrome in pregnancy and its effect on the physiologic condition.

Case Report

A 26-year-old primigravida at 11 weeks of gestation came to the outpatient department with the complaints of painful raised skin lesions associated with fever for the past 3 days. There was no history of drug intake prior to the onset of the skin lesions. Medical history was otherwise unremarkable.

Dermatological examination revealed multiple, erythematous, tender papules, nodules and plaques over both the cheeks and upper and lower limbs. Mucous membranes were normal. The differential diagnoses considered were Sweet's syndrome, urticarial vasculitis and erythema multiforme (Fig. 1).

Blood investigations revealed leukocytosis with neutrophilia and elevated ESR. All the other investigations including LFT and RFT were normal. Fetal USG revealed

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Fig. 1 Multiple ill-defined erythematous papules nodules and plaques

live fetus with no abnormality. Pathergy test was negative. Antinuclear antibodies were also negative. Skin biopsy was done, and the specimen was sent for histopathological examination.

The microscopic features of the lesional skin biopsy included normal epidermis with the dermis showing moderate-to-dense perivascular infiltration of neutrophils, lymphocytes, a few histiocytes and minimal nuclear debris indicating leukocytoclasia. There was endothelial prominence in the vessels along with minimal extravasation of erythrocytes. There was neither vessel wall destruction nor necrosis. With these features, a diagnosis of Sweet's syndrome was made (Fig. 2)

Discussion

Sweet's syndrome was first described by Dr. Robert Douglas in the year 1964 [1]. The synonyms for this disease are acute febrile neutrophilic dermatoses and Gomm-Button disease.

Sweet's syndrome can be broadly classified into three types.

1. Classical or idiopathic Sweet's syndrome,
2. Malignancy-associated Sweet's syndrome,
3. Drug-induced Sweet's syndrome [1].

Fig. 2 HPE 40×: The dermis showing moderate-to-dense perivascular infiltration of neutrophils, lymphocytes, a few histiocytes and minimal nuclear debris indicating leukocytoclasia

This was modified, and another classification of Sweet's syndrome was proposed by Von den Driesch. According to him, it is categorized into 4 groups: classic idiopathic (71 %), para inflammatory (16 %), paraneoplastic (11 %) and pregnancy related (2 %).

Classical Sweet's syndrome occurs most commonly in women between 30 and 60 years of age [1]. Female-to-male ratio is 4:1. The diagnostic criteria for classical Sweet's syndrome were proposed by Su and Liu in the year 1986 [1]. Further modifications in the criteria were done by Von den Driesch in the year 1994 [1] (Table 1). Our patient fulfilled the criteria for Sweet's syndrome.

Drugs also can induce Sweet's syndrome. The drug most commonly implicated is granulocyte colony-stimulating factor [1]. Many other drugs including antibiotics, antiepileptics, antiretroviral drugs, antihypertensives, contraceptives, diuretics, NSAIDs and retinoids have been implicated [1].

Certain malignancies are associated with Sweet's syndrome. Acute myelogenous leukemia is the most common association. Other neoplasms associated are carcinomas of the genitourinary tract, GIT, breast, ovary. There appears to be a temporal association between the onset of the skin lesions and recurrence of the neoplasm [2].

There is a myriad of clinical manifestations of Sweet's syndrome. This varies from papules, nodules, plaques commonly to bullous lesions, pustules, subcutaneous nodules and ulcerations rarely [1]. The classical histopathological feature in Sweet's syndrome is a dense infiltrate of neutrophils in upper dermis and upper dermal edema. Leukocytoclasia may be present. Pathergy test referred to as skin hypersensitivity is positive in Sweet's syndrome [1].

Not many cases of pregnancy-associated Sweet's syndrome have been reported in the literature, and the exact pathogenesis is unknown. It is postulated that a hormonal

Table 1 Diagnostic criteria of Sweet's syndrome [1]

Major criteria include:

1. Sudden onset of tender, erythematous plaques and nodules
2. Dense neutrophilic infiltration in dermis without leukocytoclastic vasculitis on histopathological examination.

Minor criteria include:

1. Fever $>38^{\circ}\text{C}$
2. Association with hematologic or visceral malignancy, inflammatory disease, gastrointestinal or upper respiratory tract infection, pregnancy, vaccination
3. Dramatic response to systemic steroids or potassium iodide
4. Abnormal laboratory values (3 of 4)
 - ESR $> 20\text{ mm/h}$
 - Raised CRP
 - WBC $> 8 \times 10^9/\text{L}$
 - Neutrophil count $>70\%$ of total WBC count

Presence of 2 major criteria and 2 of the 4 minor criteria confirms the diagnosis

origin may be implicated and elevated estrogen and progesterone levels may be responsible for the immunological changes in pregnancy-associated Sweet's syndrome. Saxena et al. have reported a 30-year-old woman in the second trimester of pregnancy with Sweet's syndrome. Spontaneous resolution of skin lesions was observed in this patient [2]. Mayra et al. [3] in their retrospective study of 73 cases have reported only a single case of Sweet's syndrome in association with pregnancy.

Hussain et al. [4] have reported a 33-year-old woman in her 16th week of gestation with typical manifestations of Sweet's syndrome. It is to be noted that in this patient, first pregnancy was uncomplicated with no similar skin lesions. Differential diagnosis of this condition includes vasculitis, infections and drug reactions.

Pregnancy-associated Sweet's syndrome has a good prognosis. In the majority of cases, good response to corticosteroids and even spontaneous resolution has been observed. It is to be noted that Sweet's syndrome usually does not have any effect on the outcome of pregnancy. Our patient responded to conservative management.

Conclusion

The occurrence of nodules and fever in a pregnant woman may be alarming to both the patient and the treating obstetrician. Awareness of this condition which is benign is important on the part of the treating obstetrician.

Compliance with Ethical Standards

Conflict interest None.

References

1. Cohen PR. Sweet's syndrome—a comprehensive review of an acute febrile neutrophilic dermatosis. *Orphanet J Rare Dis.* 2007;2:34.
2. Mahajan VK, Sharma NL, Sharma RC. Sweet's syndrome from an Indian perspective: a report of four cases and review of the literature. *Int J Dermatol.* 2006;45:702–8.
3. Rochael MC, Pantaleão L, Vilar EA, et al. Sweet's syndrome: study of 73 cases, emphasizing histopathological findings. *An. Bras. Dermatol.* 2011;86(4):702–7.
4. Hussain W, Craven N, Agarwal M, et al. A painful cutaneous eruption in a pregnant woman. *Clin Exp Dermatol.* 2007;32:227–8.