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Case Report

Sweet's syndrome in pregnancy: a rare case report

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ABSTRACT

Sweet's syndrome (SS) is a steroid-responsive dermatological condition characterized by fever, neutrophilia and a classical tender erythematous skin rash. Pregnancy-associated SS is an extremely rare clinical entity and must be kept in mind while approaching a case of skin rash in pregnancy. We reported a rare case of SS in pregnancy with classical features and characteristic histopathological features of neutrophilic dermatosis that had a benign course, resolving with topical steroid treatment instead of the usual systemic steroids.

Keywords: Sweet's syndrome, Pregnancy-associated Sweet's syndrome, Neutrophilic dermatosis, Topical steroid

INTRODUCTION

In 1964, Dr. Robert Douglas Sweet reported a group of 8 women suffering from an acute condition characterized by fever, leucocytosis and tender erythematous plaques over their bodies. This acute febrile neutrophilic dermatosis is now well known as SS and is characterized histologically by the presence of dense perivascular neutrophilic infiltrate with leukocytoclasis with a band-like infiltrate in the papillary dermis.[1,2] Few cases of SS have been documented. Around 2% of the confirmed cases of SS have been associated with pregnancy and tend to recur in subsequent pregnancies.[3] We hereby described the clinical features of a primigravida patient with an eventual diagnosis of SS.

CASE REPORT

A 30 year old primigravida at 13 weeks of gestation came with complaints of multiple painful skin lesions over her face for 2 days. There was a history of fever, sore throat and cough 3 days ago for which she visited her family physician, where she was prescribed personalized medications. No remarkable past or family history was noted. Medical history was otherwise not significant.

The patient was afebrile and hemodynamically stable at the time of admission. Her systemic examination was normal. Dermatological examination revealed multiple, tender, erythematous nodules and plaques with pseudovesiculations over the face, neck and upper chest region (Figure 1). The oral cavity was spared.

Per-vaginal examination revealed a 12-week size uterus with no signs of pelvic inflammatory disease. Differential diagnoses of SS, drug-associated eruptions, autoimmune disorders and erythema multiforme were made.

Blood investigations revealed a total leukocyte count of 13900/cmm with normal liver and renal function tests. C-reactive protein was 87.79 mg/liter. Skin lesional biopsy was taken and sent for histopathological examination. Urine microscopic examination showed leukocytosis and the presence of bacteria and urine culture revealed growth of *E. coli*. High vaginal swab culture and blood cultures showed no abnormal growth. Further blood tests were also done to evaluate the cause of fever and malaria, dengue and leptospirosis were ruled out. Fetal viability was confirmed by an ultrasound.

The patient was started on parenteral antibiotics along with analgesics and antipyretics. Urinary tract infection was treated with oral antibiotics. Local application of steroids was advised by a dermatologist. A clinical pharmacist's opinion was sought and adverse drug reaction as a cause could not be proved.

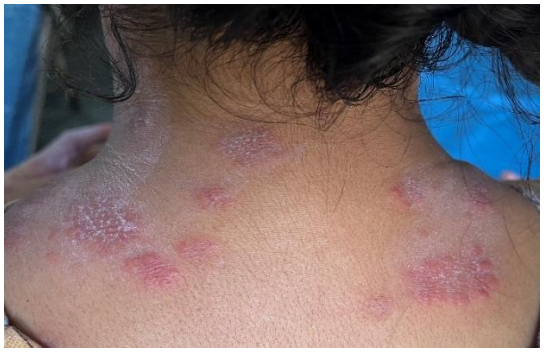

Figure 1: Skin eruptions over the neck.

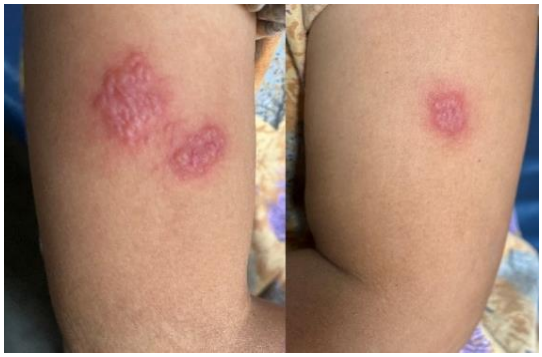

Figure 2: Tender erythematous lesions over bilateral upper limbs.

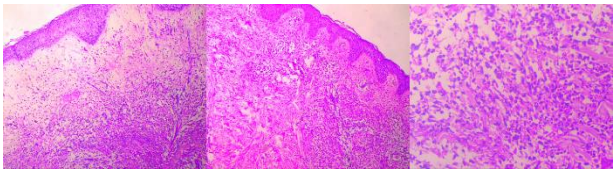

Figure 3: Histopathological examination of our patient's skin biopsy.

During the hospital stay, the patient developed similar eruptions over both upper limbs, while those on the face entered the resolution stage (Figure 2).

Multiple fever spikes were treated with antipyretics. There was a resolution of urinary and respiratory tract infections. Serial leukocyte count and CRP monitoring were done. Microscopic examination of the skin biopsy revealed epidermal acanthosis, spongiosis and exocytosis with an intrafollicular collection of neutrophils (Figure 3). Marked dermal edema with diffuse infiltrates in the upper and mid-dermis, composed predominantly of neutrophils and lymphocytes confirmed the diagnosis of SS.

DISCUSSION

SS, also known as acute febrile neutrophilic dermatosis, is a painful illness characterized by a sudden, rapid onset of fever, neutrophilia and tenderly raised erythematous skin lesions, with a diffuse dense, primarily neutrophilic infiltration in the upper dermis.[4-6]

SS can present in 3 main clinical settings: classical (idiopathic) SS; malignancy associated SS, most commonly acute myelogenous leukemia;[7] drug-induced SS first documented case post-consumption of trimethoprim-sulfamethoxazole.[8]

Classical/idiopathic (71%), parainflammatory (16%), paraneoplastic (11%) and pregnancy-related (2%) were the four basic categories developed by Dreisch.[2]

Only a few cases of pregnancy-related SS have been documented since 1964 when it was originally reported. Females between the ages of 30 and 50 were more likely to have it (male:female ratio=1:4).[9]

Table 1: Diagnostic criteria for SS.

S. no.	Major criteria	Minor criteria
1.	Sudden onset of tender erythematous nodules and plaques	Fever >38 ºC
2.	Dense neutrophilic infiltration in the dermis without leukocytoclastic vasculitis on histopathological examination	Association with hematological or visceral malignancy, inflammatory disease, gastrointestinal or upper respiratory tract infection, pregnancy, or vaccination
3.		Dramatic response to steroids or potassium iodide
4.		Abnormal laboratory values (3/4) -ESR > 20 mm/h -Raised CRP -WBC count >8×109 /l -Neutrophil count >70% of total WBC count

Su et al proposed a diagnostic criterion for SS, which was revised by Dreisch and was displayed in Table 1, requiring confirmation of two major and two minor criteria, all of which were present in our patient.[10]

The exact pathogenesis of SS in pregnancy is unknown due to a paucity of research and the disease's uncommon occurrence in pregnant women. SS is caused by an increase in proinflammatory cytokines and acute phase reactants, including G CSF and IL-6, which are increased during pregnancy.[11] Increased estrogen and progesterone levels have been proposed as a probable explanation for the symptoms, which tend to diminish following pregnancy as documented in Saxena et al case.[12]

Our patient presented with all the classical signs and symptoms that were expected in a case of SS in pregnancy such as acute onset fever and a preceding history of an upper respiratory tract infection, gastrointestinal infection or any other inflammatory condition.[2]

Dermatological picture elicited the presence of raised, tender, erythematous nodules, pustules, pseudo vesiculations and plaques, most commonly seen in the upper chest region, face and bilateral upper limbs, as in our patient. Sometimes patients can also present with bullous lesions, pustules, subepidermal nodules and ulcerations with crusting.[4] Extra cutaneous manifestations of SS were rare and infrequent including arthralgia's, blepharitis, osteomyelitis, hepatorenal failure to name a few.[4] Laboratory investigations usually revealed leukocytosis with predominant neutrophilia, thrombocytosis and elevated ESR and CRP.[2]

Histopathological examination of the skin biopsy revealed the hallmark feature of SS, a dense band-like neutrophilic infiltrate within the dermis with dermal edema, similar to that seen in our case and that reported by Hussain et al.[13] There was usually no evidence of vasculitis or fibrinoid necrosis and the presence of endothelial cell swelling, histiocytes, lymphocytes and leukocytoclasis may be predominantly seen in the later stages of the disease. The dermal edema may contribute to the pseudovesicualtions seen in some cases.[2,4]

Out of the many treatments available for SS, 30-60 mg prednisolone per day, tapered to 10 mg over 4-6 weeks had been known to work wonders and dramatic changes in lesions have been noted.[9] Local application of corticosteroid cream without systemic administration of steroids led to complete resolution of symptoms in our case. The other few documented cases of pregnancy-associated SS had also shown great responses to oral prednisolone treatments.[1] Cohen's review stated that dapsone, clofizamine and indomethacin had been effective in managing symptoms in pregnancy, however, caution should be used in patients with less than 32 weeks gestational age due to premature closure of fetal ductus arteriosus.[14]

Scope for further research

We reported the rare occurrence of pregnancy-associated sweet syndrome. Due to its incidence and reporting rarity, not much is known about the etiopathogenesis and possible role of genetics in the pathology.

CONCLUSION

We present a case of SS in a pregnant woman, which is uncommon in general and especially rarer in pregnant women. SS must always be considered during the workup of a pregnant woman with fever and skin lesions, and a skin biopsy, which is essential for diagnosis, should not be delayed. As indicated in our instance, previous accounts of SS reveal that it typically has a benign course and resolves quickly with corticosteroid therapy.

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