SKIN MANIFESTATIONS OF INTERNAL DISEASE

A CLINICAL CONUNDRUM OF NEUROSWEETS SYNDROME

P Harrison [1] - M Paul [2]

Rajagiri Hospital, Dermatology, Kochi, India [1] - Rajagiri Hospital, Hematology, Kochi, India [2]

Background: Sweet's syndrome (SS) is an inflammatory disease characterized by fever, leukocytosis and distinctive tender erythematous skin lesions that histologically consist of a diffuse dermal infiltrate of neutrophils with nuclear fragmentation. Aseptic neutrophilic inflammation may occur also in other organs. Central nervous system involvement in SS, Neuro-Sweet's syndrome (NSS), is rare. We report a case of Neuro Sweets in the setting of a hematological malignancy, coupled with a possible incriminating chemotherapeutic agent.

Observations : A 64-year-old male, with a history of coronary artery disease, admitted for dyspnoea, on evaluation, was found to have acute myeloid leukemia with spontaneous severe tumor lysis syndrome. Chemotherapy using Decitabine was initiated. In 2 weeks, he developed asymptomatic papulovesicular lesions over the forearms and face. Viral aetiology was ruled out. As the lesions progressed rapidly, he developed altered consciousness. Skin biopsy done showed neutrophilic infiltrate of dermis in the absence of vasculitis, suggestive of SS. CSF examination revealed no cells with mild protein and high sugars. All cultures were sterile. In view of deteriorating sensorium in the patient, already on high end antibiotics, antifungals and antivirals, a possibility of Neuro Sweets was considered and high dose Dexamethasone was administered. Decitabine was withheld. The cutaneous lesions and fever started resolving and Sensorium showed marginal improvement over 3 days. However, in the presence of other comorbidities, the patient eventually succumbed to multiorgan failure.

Key message: Sweet’s Syndrome an uncommon inflammatory dermatosis, though seemingly innocuous on skin, can have serious ramifications with visceral organ involvement. Neuro Sweets, a rare entity needs to be considered in encephalopathic patients with SS. This could be especially relevant when there is more than one possible trigger for this poorly understood hypersensitivity reaction. Recognition of this clinical conundrum and its poor prognosis, and an aggressive approach is warranted that could be lifesaving.