Abstract:

Background: Acute febrile neutrophilic dermatosis (Sweet's syndrome) is very rare cutaneous inflammatory disorder. It was first described by Sweet in 1964 and is the prototypic neutrophilic dermatosis and associated with infections, autoimmune and inflammatory diseases, malignancies, pregnancy and host drug exposure. Heamotological malignancies are the commonest malignancy which associated with the condition. We present a case of sweet syndrome which developed secondary to the myelofibrosis and responded well to steroid therapy.

Case Presentation: Mr N. Marapana a 62 year old male patient with previously diagnosed myelofibrosis (Jak 2 V617F negative) who was on thalidomide presented with low grade fever with flu like symptoms for 3 weeks duration and non-itchy erythematous painful ,papular ,and plaque non resolving skin rash for 3 month duration.On examination there was painful erythematous papular and nodular skin lesion over face,neck back of the chest and both upper and lower limbs.But there was no mucosal involvements. Sweet syndrome secondary to myelofibrosis, Drug eruption, connective tissue disease, paraneoplastic skin manifestation, allergic contact dermatitis and possible insect bite considered as possible differential diagnosis . His Heamoglobin 7.2 g/dl, ESR 60mm, CRP 12. ANA negative. . Repeat blood picture and bone marrow confirmed the previous diagnosis of myelofibrosis in fibrotic phase. A punch biopsy was performed and histological findings confirmed the sweet syndrome. Possible infectious and other solid and heamatological malignancies were excluded. Oral corticosteroids were started with antibiotics and there was a rapid improvement with treatment.

Conclusion: Though Sweet syndrome is very rare cutaneous inflammatory disorder, it should be considered as a differential diagnosis if a patient came with a painful erythematous papular and plaque skin lesions in appropriate clinical setting.

Key words :sweet syndrome, myelofibrosis, erythematous papular lesion