

# Sweet syndrome with differentiation syndrome related to Enasidenib

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## Abstract

Rash is frequently seen in patients with leukemia either as a sequence of thrombocytopenia, or as result of infection or allergic drug rash, however sometimes rash is pointing towards fatal complications of underlying drug therapy or underlying disease, not being aware of these alarming signs can have fatal consequences.

**Title :** Sweet syndrome with differentiation syndrome related to Enasidenib

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**Key clinical message:** Multiple conditions as Drug rash, thromboembolism, sepsis and heart failure can mimic differentiation syndrome so keeping high index of suspicion is important as the prognosis if treated is usually good and fatal if missed.

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**Figures:** 1

65-year-old female who was admitted to hospital for treatment of acute myeloid leukemia, she was on Enasidenib for IDH2+ mutation in addition to standard leukemia therapy, after one week of therapy she started having fever up to 102F with generalized skin rash, her physical exam was normal except for progressively worsening generalized skin rash as shown in figure 1A.

The initial dilemma in her diagnosis was whether this is infectious rash as she was neutropenic with absolute neutrophilic count (ANC) of 0, she was started on cefepime followed by vancomycin, the next day her rash continued to worsen with new shortness of breath, her chest x-ray showed diffuse pulmonary infiltrates

as shown in figure 1B and ANC did rapidly rise to 5 and now the clinical picture is more suggestive of differentiation syndrome with sweet syndrome.

She was a started on dexamethasone with resolution of the shortness of breath, fever, lung infiltrates and skin rash , skin biopsy was performed and it had shown prominent edema in the superficial dermis with dense infiltrate of neutrophils in the upper and mid-dermis with endothelial swelling and no evidence of vasculitis all consistent with sweet syndrome.

The main message is that differentiation syndrome happen in 20% of patients receiving isocitrate dehydrogenase inhibitor[1], it is sometimes associated with sweet syndrome, missing the diagnosis lead to fatal complications and the presentation can mimic drug rash , thromboembolism , infection or heart failure so keeping high index of suspicion is important as the prognosis if treated is usually good [1,2].

## References

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## Figure legends:

Figure 1A: showing generalized papulovesicular skin rash on the anterior abdominal wall.

Figure 1B: showing chest x-ray with evidence of bilateral diffuse interstitial lung infiltrates, no cardiomegaly.

