Cutaneous Manifestations in AML: A Case of Sweet Syndrome

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Introduction

Sweet syndrome (acute febrile neutrophilic dermatosis) is a rare inflammatory skin disorder that presents with acute onset of tender plaques/nodules, neutropenia, and fevers.

It is associated with[1]

- Infections
- Drugs
- Malignancies and autoimmune diseases

Diagnosis:[1]

- dense neutrophilic skin infiltrate without vasculitis
- fever >100.4 °F (38 °C)
- WBC >8,000/mm^3 OR >70% neutrophils)
- recent upper respiratory tract or GI infection
- malignancy
- inflammatory disease
- elevated ESR & CRP

Treatment[1]

- Discontinue potential causative drugs
- Treat underlying infection or inflammatory process
- Systemic corticosteroid

Around 4-11% of patients with Sweet syndrome are found to have acute myeloid leukemia (AML). [2,3] Meanwhile, about 1% of AML patients develop Sweet syndrome.[4]

Case Presentation

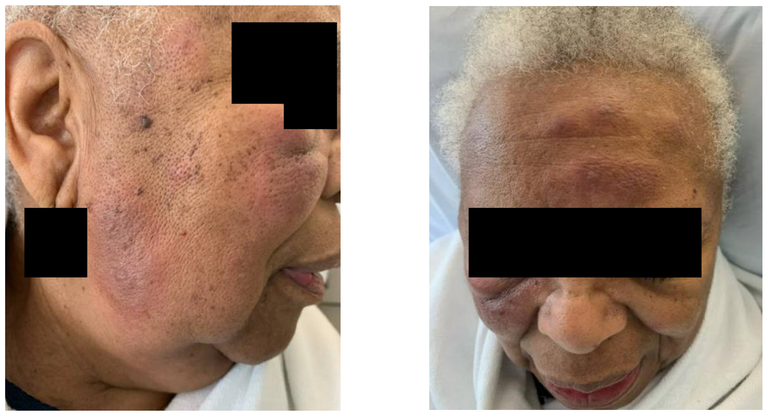

Figure 1. Inflammatory facial rash (neutrophilic dermatosis), characteristic for Sweet syndrome

85-year-old female with recently diagnosed AML, presented to the ER with anemia (Hb 6.7) and thrombocytopenia (platelets 84). Patient was admitted & started on inpatient induction chemotherapy with 5-Azacitidine and Venetoclax.

However, she began spiking persistent fevers. Extensive workup infectious workup and CT imaging did not reveal any significant findings. In the presence of recurrent fevers, new onset of a tender right-sided facial rash, and an elevated ESR/CRP, Sweet syndrome was suspected. A punch biopsy of the skin lesion was done, revealing an infiltrate of mature neutrophils accompanied by larger histiocytic appearing cells (MPO (+), CD68 (+), CD34 (-), CD117 (-), consistent with Sweet syndrome.

Steroid therapy (20-30 mg prednisone for 10 days) was started, resulting in improvements of fever curve and a gradual resolution of her facial rash.

Discussion

The pathophysiology of Sweet Syndrome is not completely understood, but it is believed to involve cytokine dysregulation causing neutrophilic infiltration into the dermis. In an underlying hematologic malignancy it may precede, coincide or present after the diagnosis is established.

A febrile rash, in the setting of known AML, should immediately prompt further investigation. Our patient fulfilled the diagnostic criteria for Sweet syndrome including characteristic febrile rash, and histopathologic findings. First line treatment involves steroid therapy which can lead to a rapid improvement of symptoms

Conclusion

This case highlights the importance of maintaining a high index of suspicion for Sweet syndrome in AML patients, who develop febrile cutaneous eruptions. Early recognition and treatment are essential to avoid complications and resolve symptoms.

References

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