“Not So ‘SWEET’” – Case of Sweet Syndrome in Patient With MDS With Excessive Blasts

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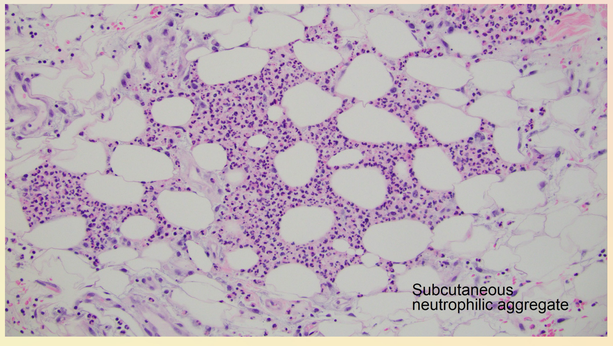
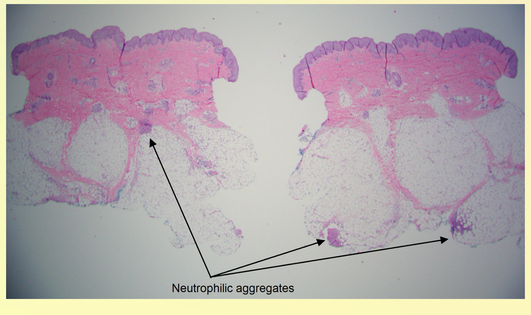
Introduction

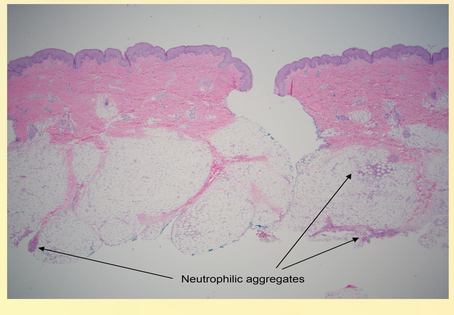
- Myelodysplastic Syndrome (MDS) is a group of disorders characterized by
  - abnormal myeloid maturation resulting in peripheral cytopenia and bone marrow dysplasia.
  - MDS with excess blasts (MDS-EB) is defined as presence of 5-19% of blasts in the peripheral blood or bone marrow and may progress to AML with blast percentage >20%.
- Sweet Syndrome (SS) is a rare inflammatory skin condition that can be secondary to chemotherapy or the underlying malignancy
  - associated with AML and MDS in adults, however, is particularly rare in children.
  - pathophysiology is thought to include hypersensitivity reactions, cytokine dysregulation especially G-CSF and genetic susceptibility to the disease process.
- Major diagnostic criteria include
  - abrupt onset of painful erythematous plaques/nodules
  - histopathologic evidence of sterile neutrophilic panniculitis.
- Minor criteria include
  - excellent response to steroids,
  - underlying malignancy
  - three of the following: ESR >20 mm/hr, CRP, >8,000 leukocytes and >70 percent neutrophils.

Case Report

- We present a case of a 4-year-old male with SS associated with MDS-EB undergoing chemotherapy.
- Patient had previously failed therapy with Azacytidine now admitted for bridge chemotherapy with cytarabine and Erwinia L-asparaginase as per modified AAML1031.
- Despite morphine, pain associated with these lesions worsened, hindering ambulation. He had similar nodules during previous induction cycles starting around his ANC nadir.
- A biopsy showed patchy predominant lobular neutrophilic panniculitis and focal neutrophilic folliculitis without malignant infiltration.







Discussion

- Laboratory results remarkable for ESR 67, CRP 302, ferritin 1,398.
- These above findings and the patient’s clinical presentation course supported the clinical diagnosis of SS.
- Given his immunosuppressive status, steroid treatment was deferred.
- Patient was treated with ketorolac and supportive care, and the lesions and pain gradually improved as his ANC counts recovered with a similar pattern to prior cycles.

Conclusion

- This atypical presentation of SS presents the first case report of a pediatric patient with SS secondary to MDS-EB.
- An abnormal response in this patient’s endogenous G-CSF production for promoting bone marrow recovery is proposed to be the trigger that led to development of SS.
- This response observed with anti-inflammatory treatment poses the possibility of considering this treatment as an alternative for pain control during the peak of immunosuppressive state while undergoing chemotherapy.

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