“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.