Intractable Pyoderma Gangrenosum in Myelodysplastic Syndromes with Isolated Trisomy 8

서울아산병원 내과[1]

위형석[1], 정지윤[1], 정정희[1], 송하림[1], *최윤숙[1]

Background: Myelodysplastic syndromes(MDS) are clonal myeloid disorders characterized by peripheral cytopenia and ineffective bone marrow hematopoiesis. MDS is known to be associated with autoimmune diseases, in which trisomy 8 is frequently found. Neutrophilic dermatosis, including Sweet syndrome and pyoderma gangrenosum(PG), has been reported to be the most frequent autoimmune manifestation.

Case: A 30-year-old man with MDS, excess blast-1(EB-1) presented with fever and a purpuric tender skin plaque on the left inguinal area(Figure 1). Neutrophil-dominant leukocytosis(10,500/uL) and elevated CRP(14.58 mg/dL) were observed. One month ago, anemia worsened with 2% of increased peripheral blasts, and disease status progressed to high-risk MDS. Cytogenetic analysis revealed trisomy 8. On the targeted whole exome sequencing, U2AF1, CBL, GNAS, and KRAS were also found, and Molecular international prognostic scoring system(IPSS) was very high. Therefore, we had planned allogeneic bone marrow transplantation. His skin lesion was abruptly developed and initially suspected as Sweet syndrome because he had a history of biopsy confirmed Sweet syndrome on deltoid muscle developed after vaccination three years ago. He was treated with 1mg/kg of methylprednisolone and empirical antibiotics. Skin punch biopsy results revealed neutrophilic dermatosis in the superficial dermis and lymphohistiocytic infiltration(Figure 2) with no evidence of infection. The morphologic finding of painful skin ulcers with undetermined borders and peripheral erythema was preferred to PG. The extent of the skin lesion and ulceration rapidly aggravated, and the steroid dose was increased to 2mg/kg/day on the 3rd day of admission. We decided to start a hypomethylating agent, decitabine, on the 4th day to control underlying MDS. Nevertheless, on the 6th day, the skin desquamation and pain rapidly worsened, and we added colchicine, cyclosporine, and indomethacin. After decitabine therapy, PG improved along with neutropenia(340 /uL) on 19th day.

Conclusion: MDS with trisomy 8 can cause neutrophilic dermatosis. Decitabine therapy can control refractory PG through bone marrow suppression.

Figure 1. Serial photography of skin lesion on the left inguinal area according to hospital day(HD)

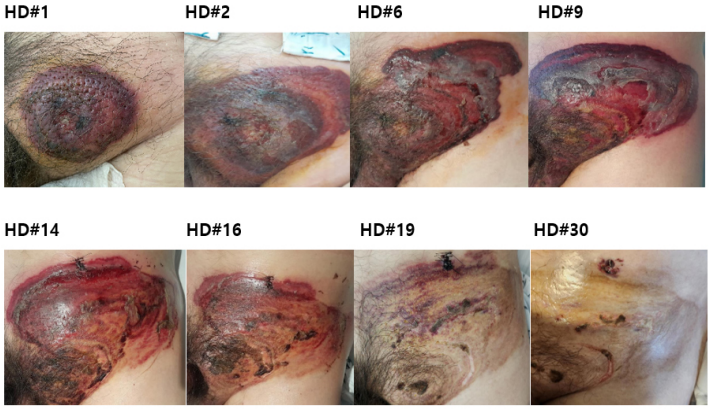

Figure 2. Pathologic findings of skin lesion on the left inguinal area.

(A) Marked neutrophil infiltration in the superficial dermis
(B) Moderate pan-dermal lymphohistiocytic infiltration