

Extranodal NK/T-cell Lymphoma, Nasal Type

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A 26-year-old male presented with a 3-month history of pain and swelling over nose without any fever, weight loss, or night sweats. Examination revealed a large necrotic mass on his nose (Figure-1a) and a perforating ulcer on the hard palate (Figure-1b). His blood counts were normal. Biopsy from the nasal lesion revealed large atypical cells with brisk mitosis. Immunohistochemistry was positive for CD2, CD3, CD7, CD56, perforin and granzyme, but negative for CD20, CD5, CD4, CD8, ALK and CD30. Epstein-Barr encoding region in-situ hybridization was positive on the biopsy, confirming the diagnosis of extranodal NK/T cell lymphoma, nasal type (ENKTCL-NT). Epstein-Barr virus DNA was detected in the peripheral blood (90900 IU/ml). PET-CT scan showed stage IV disease. Patient initially responded to SMILE chemotherapy (steroids, methotrexate, Ifosphamide, l-asparaginase and etoposide), but died later due to hemophagocytic lymphohistiocytosis. Extranodal NK/T-cell Lymphoma, nasal type is an aggressive subtype of non-hodgkin's lymphoma (NHL) caused by EBV. It is prevalent in certain Asian (China, Japan, Thailand, Korea, Taiwan) and Latin American countries (Mexico, Peru, Brazil, Guatemala).^{1,2} Unlike other NHL's, anthracycline-containing regimen are ineffective for the treatment of ENKTCL-NT. Due to its high efficacy, asparaginase backbone is cornerstone in the management of ENKTCL-NT.^{2,3}

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Running head: Extranodal NK/T-cell lymphoma

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