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A Case of Extranodal Natural Killer/T-Cell Lymphoma, Nasal Type with Dermatomyositis

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Dear Editor:

A 27-year-old male presented to our dermatology clinic with recurrent facial rash for five years, aggravated swelling for six months, and myalgia for the last two months. He presented with erythema and edema of the face (Fig. 1A). There was no rash on his trunk or limbs. Proximal dominant muscle weakness in his upper limbs (Medical Research Council grade 4) with grasping pain was noted. In addition, he complained that he had a history of rhinitis lasting 3 years.

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Laboratory investigations showed elevated creatine kinase, 1,108.2 U/L (55～170 U/L); lactate dehydrogenase, 1,283 U/L (98～192 U/L); alanine transaminase, 74.2 U/L (9～50 U/L); aspartate transaminase, 85.5 U/L (15～40 U/L); erythrocyte sedimentation rate, 19 mm/h (0～15 mm/h). Needle electromyography revealed fibrillation potentials, positive sharp waves, short duration low amplitude motor unit potential with increased polyphasic potential, and early recruitment in proximal muscles of the upper limb. A muscle biopsy of left bicep brachii on frozen sections suggested inflammatory myopathy (Fig. 1B). The right mandible skin lesions biopsy revealed epidermal focal parakeratosis, and liquefaction degeneration of basal cells, lymphocytic cell infiltration with mild atypia around vascular regions, appendages and in the fat lobules (Fig. 1C～E). Immunohistochemistry analysis showed positive of Ki67 (30%), LCA, CD2, CD3, CD8, CD4, TIA-1, and Epstein-Barr encoding region (EBER); negative of CD79a, CD20, CD3; weak positive of CD56 and granzyme B (Fig. 1F～J). We suspected that it was extranodal natural killer (NK)/T-cell lymphoma, nasal type (ENKTL) but not sure, then the patient was admitted for study and treatment. After admission the patient developed a high fever, pharyngalgia, and dysphagia. A nasopharyngeal biopsy was performed and showed massive cells infiltration which were stained positive for cytoplasmic CD3, CD56, EBER (by *in situ* hybridization), granzyme B, TIA-1, and Ki67 (about 50%) with

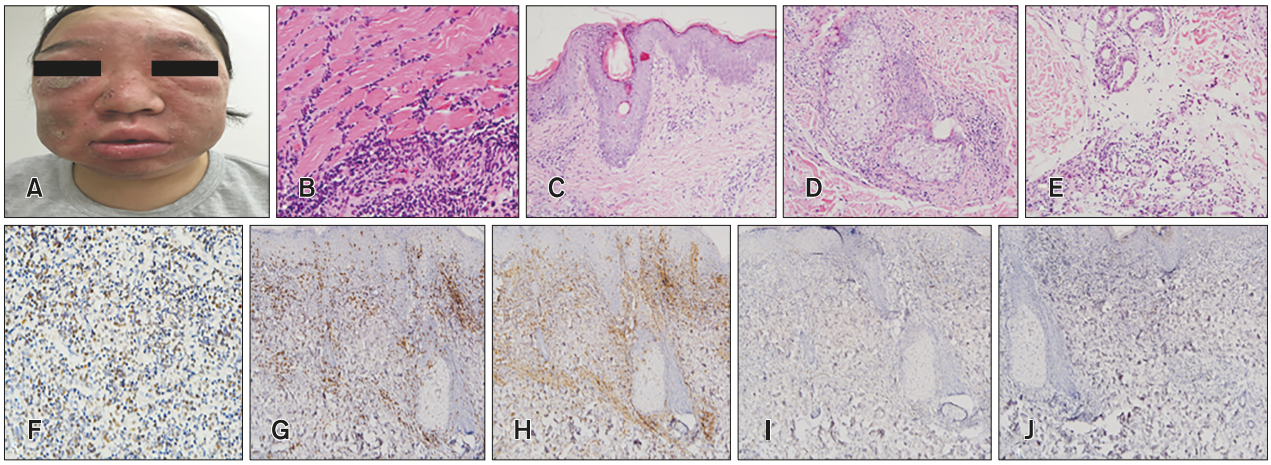

Fig. 1. (A) The patient with erythema, erosive lesions, crusts and edema on the face. (B) Skeletal muscle fascicles revealing markedly distorted overall architecture with inflammatory interstitial infiltrate of lymphocytes and perimysial expansion without atypia (H&E, ×400). (C~E) Histopathology analysis of right mandible skin lesions revealed epidermal focal parakeratosis and liquefaction degeneration of basal cells, lymphocytic cell infiltration with mild atypia around vascular regions, appendages and in the fat lobules (H&E, ×200). Immunohistochemistry analysis (F: ×400; G~J: ×200) showed Epstein-Barr encoding region (EBER) (F), CD3 (G), and CD4 (H) were positive; CD56 (I) and granzyme B (J) were weak positive. We received the patient's consent form about publishing all photographic materials.

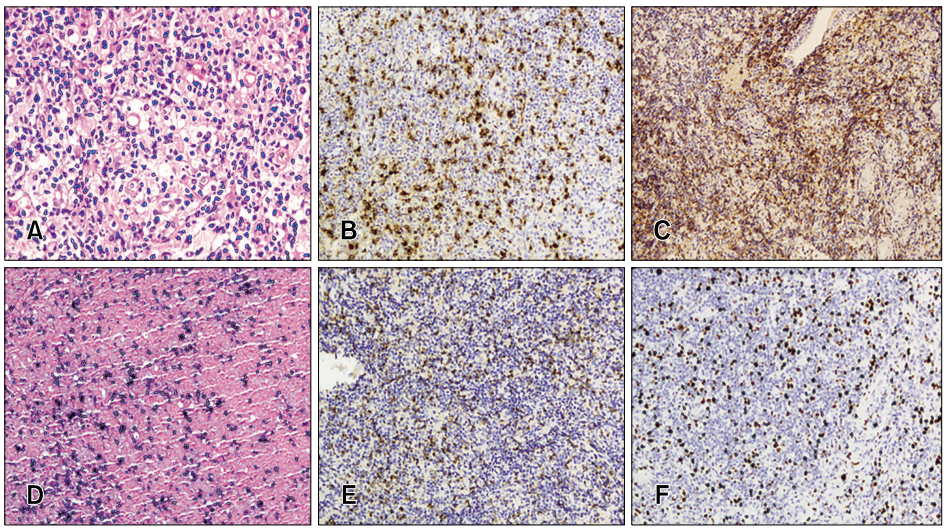

Fig. 2. (A) A nasopharyngeal biopsy showed massive cell infiltration with atypia (H&E, ×400). Immunohistochemistry analysis (×400) showed positive for granzyme B (B), CD56 (C), Epstein-Barr encoding region (EBER) (by *in situ* hybridization) (D), TIA-1 (E), and Ki67 (about 50%) (F).

atypia (Fig. 2). These established the diagnosis of ENKTL with dermatomyositis. Bone marrow puncture examination showed no abnormality. However, he and his family gave up treatment and left the hospital, then died after 10 days because of deterioration of the disease and hemophagocytic syndrome.

Nasal ENKTL is a mature form of T/NK-cell lymphoma and have strong associations with Epstein-Barr virus[1]. ENKTL is the most common type of nasal lymphoma in Asia and central America[2]. The typical immunophenotype of ENKTL is CD56+ and cytoplasmic CD3+. CD56 is a neuronal cell adhesion factor that promotes the adhesion of tumor cells to the walls of blood vessels, causing significant features of vascular destructive infiltration. In addition, the most common hematological malignancies associated with dermatomyositis are B-cell lymphomas, but they are rarely associated with T-cell lymphoma and NK/T-cell lymphoma[3]. When reviewing the literatures, we found two cases of paraneoplastic phenomena were described as dermatomyositis/myositis[4,5] as our case. ENKTL, nasal lymphoma is rarely associated with skin lesions mimicking inflammatory or reactive disorders with a similar histopathologic pattern.

In conclusion, our case highlights the association of paraneoplastic disorders such as dermatomyositis with ENKTL which is difficult to be appropriately diagnosed based on skin and muscle lesions in the early stages. So it needs us

to follow-up for a long time, especially in patients with rhinitis.

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CONFLICTS OF INTEREST

The authors have nothing to disclose.

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DATA SHARING STATEMENT

Research data are not shared.

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