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Huge Steatocystoma Multiplex with New Point Mutation in the Exon 1 of KRT 17 Gene

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Dear Editor:

A 50-year-old woman presented with huge multiple cysts on the back, chest, abdomen, neck, axillae, antecubital fossae, and popliteal fossae for 40 years (Fig. 1A). All of the cysts were asymptomatic and non-inflammatory. Her father and her son also had cysts of same nature. However, the cysts were not huge in size. Her laboratory findings, including CBC, urinalysis, VDRL, LFT/RFT, PT/PTT, and HB markers, were normal or within normal limits. Histopathologic findings showed a huge cyst surrounded by stratified squamous epithelium with adjacent sebaceous glands (Fig. 2A). Immunohistochemistry using CK-17 antibody was positive on the cyst wall and sebaceous glands (Fig. 2B). She was diagnosed with steatocystoma multiplex. We carried out mutation analysis of KRT 17 gene using direct sequencing. We found new point mutation in the exon 1 of KRT 17 gene (c. 425G>T) (Fig. 2C). She underwent operations several times and finally got a good cosmetic result (Fig. 1B). We received patient consent form for publishing photos.

Steatocystoma multiplex is a rare disorder manifested as skin-colored subcutaneous cysts[1]. It usually begins on the chest and abdomen in pubertal period, and varies from matchhead to bean in size. However, the cysts in this case were extraordinarily huge. In Korea, only Jeong et al.[2] reported giant steatocystoma multiplex limited in the scalp. Pathogenetically, steatocystoma multiplex is usually associated with mutation in the KRT 17 gene exhibiting autosomal dominance[1]. Keratin 17 is a type I cytokeratin which is found in nail beds, hair follicles and sebaceous glands. Based on the data of several studies, different mutations can develop the same clinical phenotype in steatocystoma multiplex, and the same mutations can cause different clinical phenotypes[3]. Therefore, it is supported that

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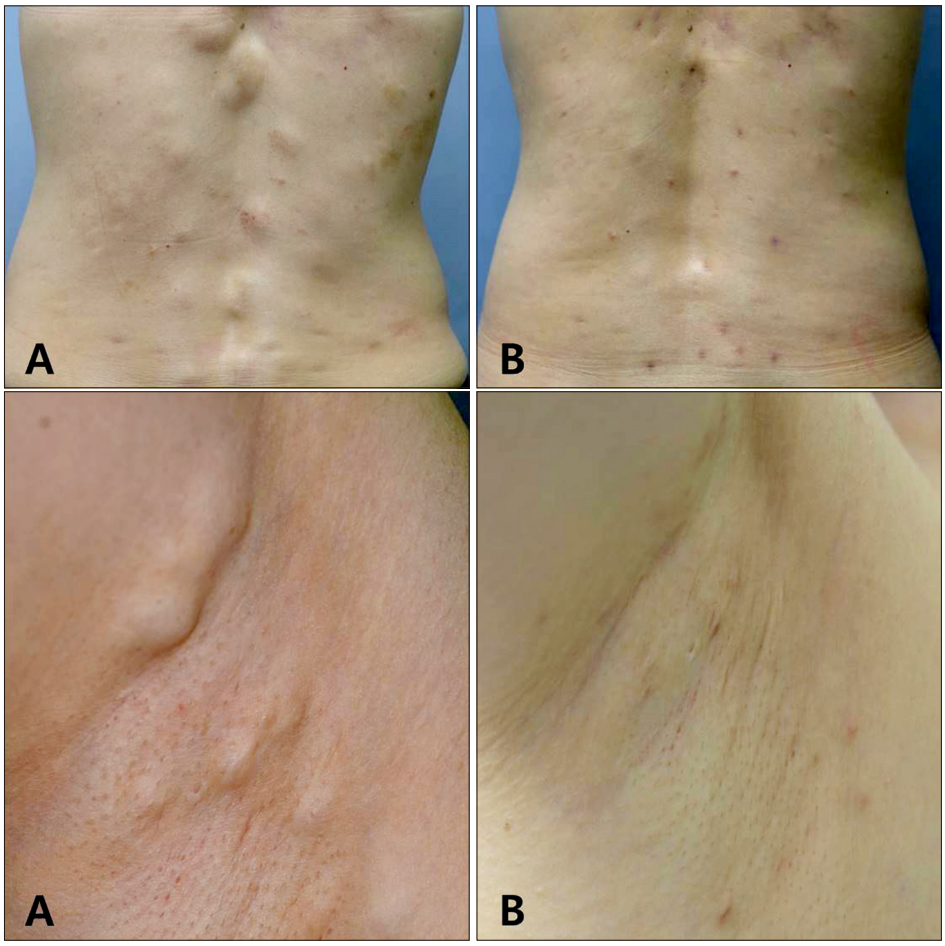

Fig. 1. (A) Huge multiple cysts on her back and right axilla and (B) improved status after operation.

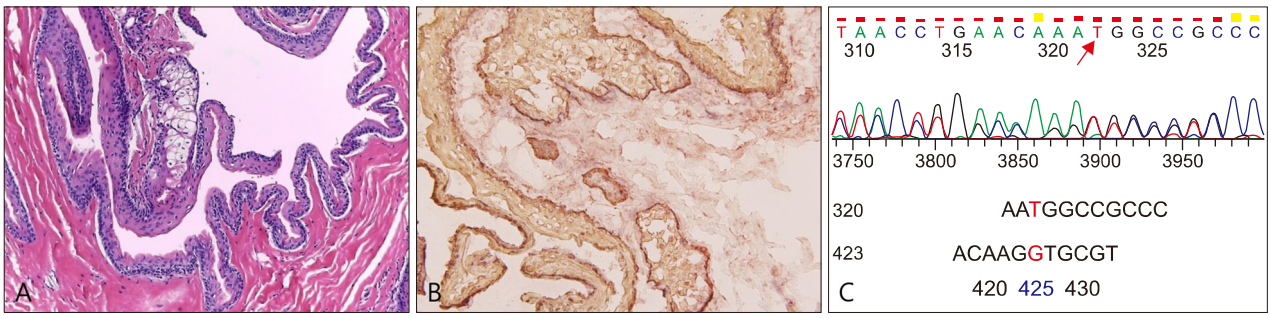

Fig. 2. (A) A cyst covered by stratified squamous cells with adjacent sebaceous glands (H&E, ×100). (B) Immunohistochemistry was positive on the cyst wall and sebaceous glands (CK-17, ×200). (C) Missense mutation in exon 1 of KRT 17 gene (c. 425G>T).

the genotype-phenotype correlation of steatocytoma multiplex can be determined, not only by the KRT 17 gene mutation but also other modifying factors including the site, androgenic stimulation and environmental factors[4,5]. Although this case had huge steatocytoma multiplex with point mutation in the exon 1 of KRT 17 gene, the correlation between genotype and phenotype should be determined. The various modalities in the treatment of steatocystoma multiplex include surgery, laser therapy, cryotherapy, radiofrequency incision probe and oral isotretinoin1. This patient was treated with surgical procedure and got a good cosmetic result. We here report a case of huge steatocystoma multiplex with new point mutation in the exon 1 of KRT 17 gene.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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A Case of Mycosis Fungoides Occurring after Primary Gamma-Delta T-Cell Lymphoma

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Dear Editor:

Mycosis fungoides (MF) represents the most common type of cutaneous T-cell lymphoma, characterized by epidermotropic proliferation of small-to medium-sized T-cells[1]. On the other hand, primary cutaneous gamma-delta T-cell lymphoma (PCGD-TCL) is composed of a clonal proliferation of mature, activated gamma-delta T-cells with a cytotoxic phenotype[1]. Here, we report a case of MF following PCGD-TCL. To our knowledge, this is the first reported case.

A 52-year-old woman presented with a 2-year history of erythematous lesion with crust and erosion on the right pubic area. She had no particular medical history. A biopsy specimen revealed infiltration of medium to large sized atypical lymphoid cells in the dermis and subcutis (Fig. 1). We received the patient's consent form about publishing all photographic materials. The majority of atypical cells were CD3+, CD4−, CD8−, and CD56+. Also immunostaining revealed the absence of T-cell receptor (TCR)-βF1. Laboratory examination revealed leukocytosis (9,900/μl; normal: 3,150∼8,630/μl). Positron emission tomography showed no internal organ involvement. The patient was diagnosed with PCGD-TCL. The patient received multidrug chemotherapy; 6 cycles of bortezomib, cyclophosphamide, adriamycin, vincristine, and prednisolone. After 1 year PCGD-TCL recurred and the patient received radiotherapy after 6 cycles of chemotherapy. This resulted in complete response for PCGD-TCL. Three years later, the patient revisited with erythematous scaly patches on her extremities and trunk. A biopsy specimen showed epidermal and dermal infiltration of atypical lymphocyte (Fig. 2). Phenotypes of these cells were CD3+, CD4+,

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