Two Cases of Diffuse Plane Xanthoma

Jung-Chul Choi, M.D., Sung-Eun Chang, M.D., Jee-Ho Choi, M.D.,
Kyung-Jeh Sung, M.D., Kee-Chan Moon, M.D., Jai-Kyoung Koh, M.D.

Department of Dermatology, Asan Medical Center, College of Medicine, University of Ulsan, Seoul, Korea

Diffuse plane xanthoma characteristically presents with asymptomatic, yellow-orange plaques that usually involve periorbital areas, neck, upper trunk and flexural folds. Although most cases of diffuse plane xanthoma are associated with multiple myeloma, several cases have been described with different lymphoproliferative disorders and other miscellaneous diseases. We report herein two cases of diffuse plane xanthoma which are not associated with underlying disease.(Ann Dermatol 13(3) 179~182, 2001).

Key Words : Diffuse plane xanthoma

Diffuse normolipemic plane xanthoma(DPX) was first described by Altman and Winkelmann in 1962[1]. It characteristically presents with asymptomatic, yellow-orange plaques that usually involve periorbital areas, neck, upper trunk and flexural folds. Histopathologically, the lesions of DPX show single arrangement, small clusters, and large sheets of foam cells, diffusely scattered throughout the dermis. Initially DPX was not believed to be associated with internal diseases. In 1966, Lynch and Winkelmann recognized the relationship of DPX to diseases of the reticuloendothelial system[2]. Although most cases of DPX are associated with multiple myeloma[3], several cases have been described with different lymphoproliferative disorders and other miscellaneous diseases[4-12]. In Korea, four cases of DPX have been reported[13-15]. Only one case[13] showed hyperlipoproteinemia and two cases[13-14] were not associated with underlying disorders. We report herein a case of DPX which shows normolipoproteinemia and is not associated with underlying disease, and a case of DPX which shows hyperlipoproteinemia and is not associated with underlying disease.

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Reprint request to : Kee-Chan Moon, M.D. Department of Dermatology, Asan Medical Center, College of Medicine, University of Ulsan, 388-1, Poongnap-Dong, Songpa-Ku, Seoul, 138-736, Korea
Tel. 2224-3460, Fax. 486-7831
e-mail. derm@www.amc.seoul.kr

CASE REPORT

Case 1.

A 57-year-old Korean woman presented with a 2-month history of asymptomatic, yellow-orange colored plaques on the periorbital area, right preauricular area and neck. There was no history of preceding disorders and inflammation. She had no family history of hyperlipemia. She was not taking any oral medications. Skin examination revealed several, 1.0 to 2.0cm, well-defined, irregular, yellow-orange plaques involving the periorbital area, right preauricular area (Fig. 1) and neck. A complete blood cell count, erythrocyte sedimentation rate, liver function test, and urinalysis were within normal limits. Serum lipids showed a total cholesterol of 236 mg/dl (130-240 mg/dl) with a high-density lipoprotein (HDL) fraction of 58 mg/dl (45-65 mg/dl), low-density lipoprotein (LDL) fraction of 150 mg/dl (95-215 mg/dl), and triglyceride level of 139 mg/dl (0-200 mg/dl). A skin biopsy specimen taken from the preauricular area showed clusters of foam cells, as well as foam cells singly, diffusely scattered throughout the dermis (Fig. 2). Multinucleated giant cells were seen. Occasionally a perivascular arrangement of foam cells was noted. No evidence of

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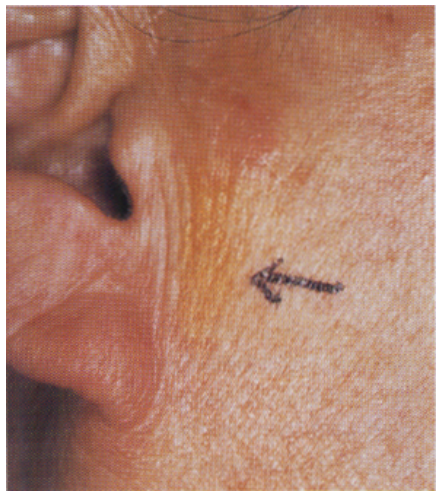

Fig. 1. Case 1. A well-defined 2.0 × 1.0 cm yellow-orange plaque on the right preauricular area.

necrobiosis was present. The fat stains were positive, and the clinicopathological findings were interpreted as consistent with diffuse plane xanthoma. The patient has been followed up without treatment for 6 months.

Case 2.

A 65-year-old Korean woman presented with a 1-year history of asymptomatic, yellow-orange colored flat papules on the periorbital area, neck and chest. There was no history of preceding disorders and inflammation. She was not taking any oral medications and was otherwise healthy. Skin examination revealed several, 0.5 to 1.0cm, irregular, yellow-orange flat papules involving the periorbital area, neck and upper chest (Fig. 3). Serum

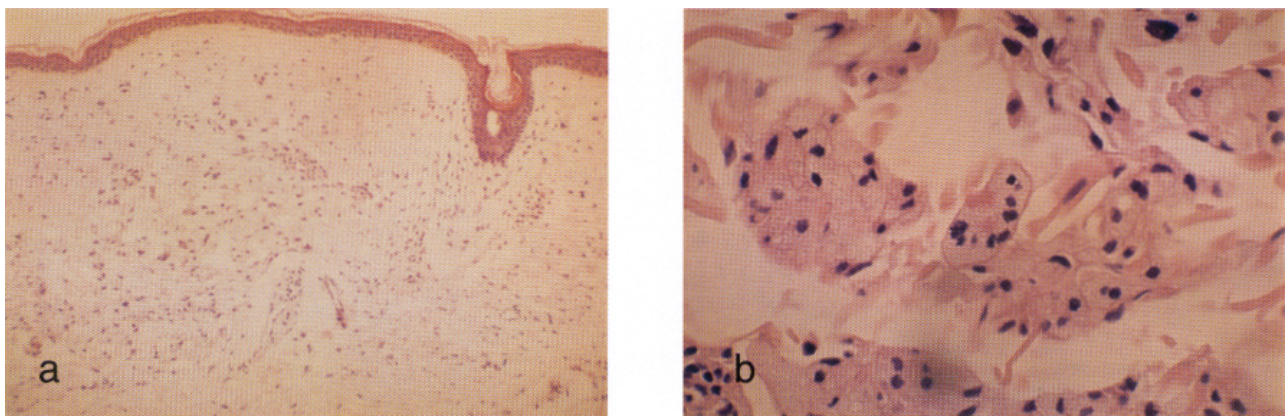

Fig. 2. Case 1. (a) A skin biopsy specimen taken from the right preauricular area showed the clusters of foam cells, diffusely scattered throughout the dermis (H&E stain, ×100). (b) Multinucleated giant cells were also seen (H&E stain, ×400).

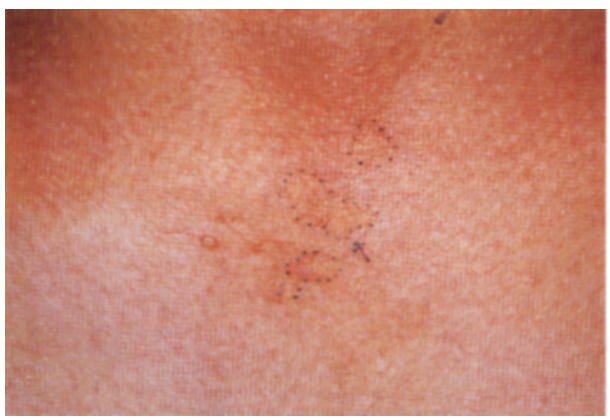

Fig. 3. Case 2. Several 0.5 to 1.0cm yellow-orange flat papules on the neck and chest.

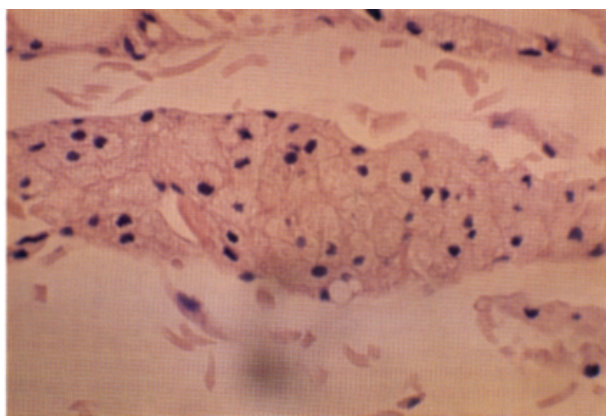

Fig. 4. Case 2. Large sheets and clusters of foam cells in the dermis (H&E stain, ×400).

lipids showed a total cholesterol of 264 mg/dl with a high-density lipoprotein (HDL) fraction of 58 mg/dl, low-density lipoprotein (LDL) fraction of 184 mg/dl, and triglyceride level of 111 mg/dl. A skin biopsy specimen taken from the chest showed large sheets and clusters of foam cells, diffusely scattered throughout the dermis (Fig. 4). Multinucleated giant cells were also seen. The lesions were diagnosed as diffuse plane xanthoma. The patient has been treated with simvastatin (5mg/day) and low fat diet for 5 months. The lesion remained stable without improvement.

DISCUSSION

Diffuse normolipemic plane xanthoma was first described by Altman and Winkelmann in 1962[1]. It characteristically presents with asymptomatic, yellow-orange plaques that usually involve periorbital areas, neck, upper trunk and flexural folds. In 1966, Lynch and Winkelmann recognized the relationship of DPX to diseases of the reticuloendothelial system[2]. Although most cases of DPX are associated with multiple myeloma[3], several cases have been described with different lymphoproliferative disorders including benign monoclonal gammopathy[4], macro- globulinemia[5], cryoglobulinaemia[6], leukemia[7], lymphoma[8], mycosis fungoides[9], and Castleman's disease[10]. Other reported associated diseases include erythroderma[11], and Ehlers-Danlos syndrome[12].

Three categories of normocholesterolemic xanthomatosis are generally accepted[16-17]. Type I includes a group of patients with alteration of structure or content of lipoproteins. Type IA is associated with the accumulation of lipids within lipoproteins other than cholesterol such as cholestanol and phytosterols. These rare genetic disorders are characterized by cerebrotendinous xanthomatosis having xanthomas in the brain, lung, and tendons, and by phytosterolemia characterized by increased absoption of plant sterols with the tendinous and tuberous xanthomas. Type IB represents a group of disorders characterized by alteration in the lipoprotein. Dysbetalipoproteinemia, HDL deficiency, and serum lipoprotein deficiency are included within this type. Type II is associated with lymphoproliferative diseases. Type III includes a group of patients with clinical and histologic findings of normocholesterolemic xanthomas but without underlying lymphoproliferative disease or lipoproteins abnormality[18].

In one study[19], only 3 of the 8 DPX patients had an associated lymphoproliferative disorder. These 3 cases had disseminated lesions involving trunk and extremities, suggesting that an extensive cutaneous involvement may indicate the presence of an associated systemic disease. In Korea, four cases of DPX have been reported[13-15]. One patient[13] presented with yellowish plaques on the periorbital area, neck and upper chest. She had no underlying systemic disorders. Three patients[14-15] presented with yellowish patches on the face, neck, trunk and extremities, of which two patients were associated with monoclonal gammopathy. Our patients presented yellow-orange flat papules and plaques on the face, neck and chest clinically, and had no underlying disease. However, our cases can not preclude that underlying diseases may develop because the follow up period is not so long.

Histopathologically, the lesions of DPX shows large sheets and clusters of foam cells, as well as foam cells singly and in small groups, diffusely scattered throughout the dermis[20]. In some areas, the foam cells lie in thin streaks between collagen bundles, and occasionally a perivascular arrangement is noted. There may be an admixture of histiocytes and lymphoid cells. Rarely, Touton giant cells are seen. Our patients showed foam cells scattered throughout the dermis and multinucleated giant cells.

The differential diagnosis includes xanthoma disseminatum and necrobiotic xanthogranuloma. Xanthoma disseminatum is a rare condition which consists of a triad of papular/planar xanthomatous lesions involving the flexures and intertriginous areas, mild transient diabetes insipidus in up to 40% of patients, and xanthomata of the mucous membranes of the upper respiratory tract in some cases[21]. Necrobiotic xanthogranuloma has a characteristic histological picture, with confluent granulomatous masses seen as either sheets or nodules, replacing most of the dermis, and extending to the subcutaneous tissues with numerous giant cells[22].

When a DPX patient has an underlying disorder, it should be treated. The DPX patient with hyperlipoproteinemia is treated with low fat diet, cholestyramine, fibrates, probucol, which have been unsuccessful in resolving the xanthoma[23]. Our patient (case 2) has been treated with simvas-

tatin (5mg/day) and low fat diet for 5 months. However, the lesion remained stable without improvement. In a DPX patient without any associated systemic disease, treatment is not needed except for cosmetic purpose.

We report a case of DPX which shows normolipoproteinemia and is not associated with underlying disease, and a case of DPX which shows hyperlipoproteinemia and is not associated with underlying disease.

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