A Case of a Subcutaneous Juvenile Xanthogranuloma

Jeong Deuk Lee, M.D., Chung Eui You, M.D., Chang Nam Lee, M.D., Hoon Kang, M.D.*, Sang Hyun Cho, M.D.

Department of Dermatology, College of Medicine, The Catholic University of Korea, Seoul, Korea ChoiKang Skin and laser clinic, Seoul, Korea*

Juvenile xanthogranuloma is a congenital or perinatal tumor, 1 to 2 cm in diameter, usually located on the head. The extracutaneous lesions can occur on the eye, the lung, the epicardium, the oral cavity or the testicles. Subcutaneous form of juvenile xanthogranuloma has been reported very rarely in the literature. We report a unique case of a subcutaneous juvenile xanthogranuloma that showed 4×4 cm sized plaque and located on the extremity of 9-year-old girl. (Ann Dermatol 15(1) 31~33, 2003).

Key Words : Subcutaneous juvenile xanthogranuloma

Juvenile xanthogranuloma is a benign, self-healing disorder of infants, children and occasionally adults. It is characterized by yellowish papular or nodular lesions on the skin. The extracutaneous lesions can occur rarely on the eye, the lung, the epicardium, the oral cavity or the testicles. Histopathologically mature lesions contain foamy cells, foreign body giant cells, and Touton type giant cells, which are distributed mainly in the superficial dermis and at the border of infiltration. The lesion of subcutaneous form is located in the deep dermis and the subcutis, while sparing the superficial dermis. The subcutaneous juvenile xanthogranuloma is usually a solitary deep-seated tumor sized 1 to 2 cm in diameter and is developed congenitally or during perinatal period, usually on the head. The subcutaneous juvenile xanthogranuloma has been reported very rarely in the literature. We herein report a unique case of a subcutaneous juvenile xanthogranuloma, which showed 4×4 cm sized plaque and located on the extremity.

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Reprint request to : Sang Hyun Cho M.D., Department of Dermatology, Our Lady of Mercy Hospital, College of Medicine, The Catholic University of Korea, 665, Pupyongdong, Pupyonggu, Inchon, 403-720, Korea
Tel. 82-32-510-5528, Fax: 82-32-510-5827
E-mail. drchosh@hotmail.com

CASE REPORT

A 9-year-old girl presented with a skin-colored, subcutaneous plaque on the right elbow for 1 year(Fig.1). Physical examination revealed a 4×4 cm sized, firm, movable, well-demarcated subcutaneous nodule. The patient was in good general health and the physical and laboratory examination disclosed no abnormality. The presumptive clinical diagnoses were lipoma, lymphangioma and xanthogranuloma. Histopathological examination revealed the subcutaneous nodules, which located within the reticular dermis and the subcutis, and those were well delimited but not encapsulated (Fig. 2 A). At higher magnification, the tumor was composed of foamy cells, scalloped cells, oncocytic cells and Touton type giant cells(Fig. 2 B). This specimen showed negative reactions to S-100 protein. Her parents refused complete resection of the mass and has been treated with intralesional steroid injection.

DISCUSSION

Helwig and Hackney[1] proposed the term 'juvenile xanthogranuloma' in 1954. Juvenile xanthogranuloma is the most common xanthomatous lesion occurring in infancy and considered to represent a lesion originating from histiocytes[2]. It is characterized by cutaneous papules or nodules located on the head,

neck, or extremities[3]. The disease is usually self-limited and undergoes spontaneous involution with rare exception, and usually affects both sexes equally. The serum lipid abnormalities are not accompanied[1,2]. The histopathological finding is the diffuse, uniform population of epithelioid cells admixed with Touton giant cells, eosinophils, and plasma cells, in varying proportions[2,3].

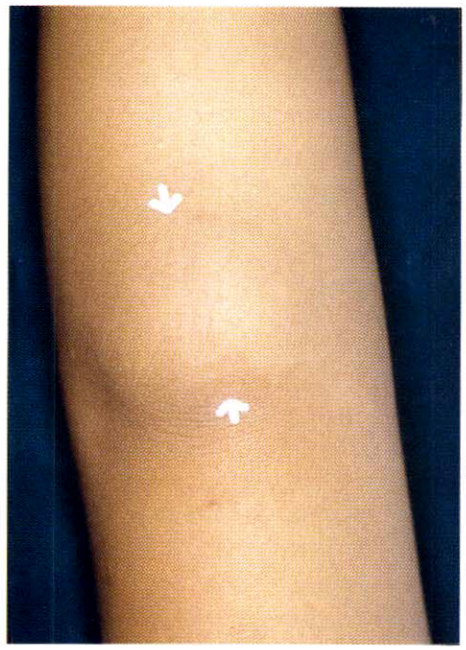

Fig. 1. A skin-colored, 4×4 cm sized, firm, movable, dome-shaped subcutaneous nodular mass on the right elbow in a 9-year-old girl.

Rarely it is located on extracutaneous sites such as subcutis, muscles, eye, lung, epicardium, oral cavity, testicles, and omentums[4,5]. There are only a few reported cases of juvenile xanthogranuloma with deeper localizations[2,5,6]. The subcutaneous juvenile xanthogranuloma was first described by Janney et al[2]. They reported 2 cases of solitary subcutaneous juvenile xanthogranuloma on the scalp and on the forehead. The sizes of the lesions were 1.2 and 2 cm respectively. De Graaf et al. reported a case of solitary, 3 cm sized, subcutaneous juvenile xanthogranuloma which occurred on the left ear[5]. In addition Sanchez et al. also reported a case of solitary, 3 cm sized, subcutaneous juvenile xanthogranuloma which occurred in the right retroauricular area[6]. The frequency of deep juvenile xanthogranuloma occurring in deep soft tissue or parenchymal or-

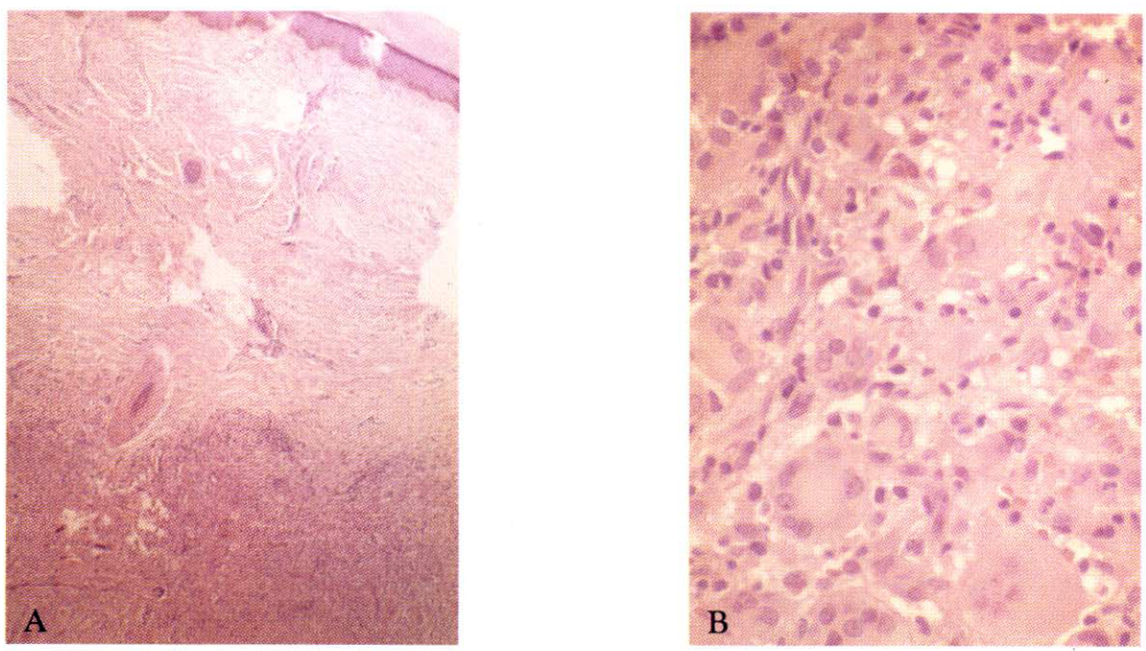

Fig. 2. (A) A well-delimited subcutaneous nodular mass within the reticular dermis and the subcutis (hematoxylin-eosin stain ×40). (B) The mixed infiltration of foamy cells, scalloped cells, oncocytic cells and Touton type giant cells (hematoxylin-eosin stain ×400).

gans has been estimated as 5%[7,8]. In comparison with the control cases of cutaneous juvenile xanthogranuloma, the deep form showed several minor histopathological differences such as prominent circumscription, fewer Touton-type giant cells, and abundant eosinophils[2,3]. The subcutaneous juvenile xanthogranuloma is characterized by solitary 1 to 3 cm sized tumor that are usually located on the head. However, our case showed a large 4×4 cm sized plaque located on the extremity. Our patient presents a unique clinical feature of subcutaneous juvenile xanthogranuloma in comparison with reported cases in the literature. The juvenile xanthogranulomas must be differentiated from histiocytosis X because the prognosis is different[4,7]. Touton giant cells, a feature of juvenile xanthogranuloma, are typically absent in histiocytosis X[4]. Immunohistochemically, histiocytosis X cells show a strong staining for S-100 protein. The tumor cells of juvenile xanthogranuloma, however, revealed no staining[7]. Juvenile xanthogranulomas, especially those located in the deep tissues, may be mistaken for malignant tumors such as embryonal rhabdomyosarcoma or as malignant fibrous histiocytoma. The usual lack of nuclear atypism and pleomorphism of the tumor cells and the absence of rhabdomyoblasts will make for a correct diagnosis[7]. In conclusion, the subcutaneous juvenile xanthogranuloma can be presented with diverse clinical features in its size, and location.

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