An Unusual Juvenile Xanthogranuloma on a Finger MCP Joint

Sang Hee Cha, M.D., Sang Hyun Cho, M.D., Jeong Deuk Lee, M.D.

Department of Dermatology, College of Medicine, The Catholic University of Korea, Seoul, Korea

Juvenile xanthogranuloma (JXG) is a benign self-limited histiocytic proliferative disorder that usually occurs in early childhood. JXG appears as reddish to yellow, papules, or nodules, and although the head, neck, and trunk are the most frequent locations, it can occur at any body site. However, JXG involving the finger is rare. Histologically, JXG is characterized by an ill-defined, unencapsulated, dense histiocytic infiltrate within the dermis, some of which is contained in Touton giant cells, foreign body giant cells and foamy cells. Because the cutaneous lesions spontaneously regress, treatment is not usually indicated. The authors report a case of JXG in a 4-year-old girl who had tender, yellowish papule on the ventral aspect of the MCP joint of the right fourth finger consistent with JXG.
(Ann Dermatol (Seoul) 20(4) 200～203, 2008)

Key Words: Finger, Juvenile xanthogranuloma

INTRODUCTION

Juvenile xanthogranuloma (JXG) is a benign cutaneous histiocytic proliferation, and was first described by Helwig and Hackney in 1954[1]. The pathophysiology of JXG is not well understood, although it is thought to originate from a histiocytic granulomatous reaction[2,3].

The cells of JXG originate from the monocyte-macrophage lineage, which can differentiate in diverse directions[2]. JXG normally occurs in infancy or early childhood[4,5], and clinically usually manifests as yellowish or red-brown, firm papules, or nodules[6]. Lesions usually present on the head, neck, and trunk, however JXG involving a finger is rare[4]; only six cases of JXG of the fingers have been reported in the English literature.

Received December 17, 2007
Accepted for publication April 16, 2008
Reprint request to: Jeong Deuk Lee, M.D., Department of Dermatology, Our Lady of Mercy Hospital, College of Medicine, The Catholic University of Korea, 665, Bupyeong-dong, Bupyeong-gu, Incheon 403-720, Korea. Tel: 82-32-510-5528, Fax: 82-32-506-9514, E-mail: leejd@olmh.cuk.ac.kr

CASE REPORT

A 4-year-old girl presented with a papule of several months duration on the ventral aspect of the right fourth finger MCP joint (Fig. 1). The lesion was a firm, dome-shaped, yellowish, 0.4×0.4 cm sized papule. There was no remarkable past or family history. On physical examination, there was

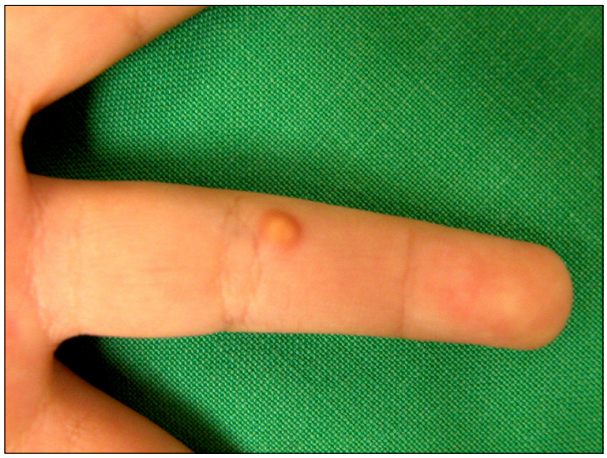

Fig. 1. A tender, solitary, firm, dome-shaped yellowish papule on the ventral aspect of the right forth finger over the MCP joint.

no limitation of joint motion and no abnormal findings other than the cutaneous lesion. A 4 mm punch biopsy specimen of the lesion showed dense intradermal histiocytic infiltrates, some of which contained foamy cells, Touton giant cells, and foreign body giant cells (Fig. 2). Scattered lymphocytes and eosinophils were also presented. Histopathological findings were consistent with a diagnosis of JXG. The papule was removed under local anesthesia using a 4 mm punch.

DISCUSSION

JXG is the most common form of non-Langerhans cell histiocytosis and is considered a benign histio-

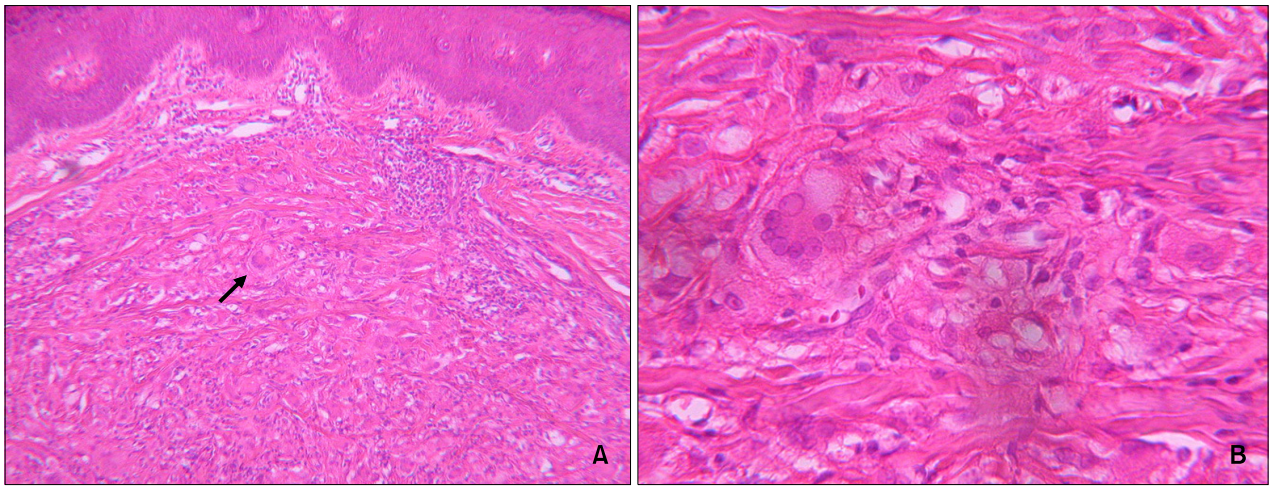

Fig. 2. (A) Dense histiocytic infiltrate in the dermis including Touton giant cells (arrow) (H&E, ×100). (B) A typical Touton giant cell and a foreign body giant cell (H&E, ×400).

Table 1. Cases of juvenile xanthogranuloma of the fingers published in the English literature

Reference	Sex/Age	Onset	Location	Associated condition	Treatment	Outcome
Sonoda et al[1]	F/27 years	2 months of age	Finger	Not stated	Not stated	Not stated
Piraccini el al[3]	M/18 months	Over 7 months of age	Proximal nail fold of right thumbnail	Severe depressed nail plate and hyperkeratotic cuticle	None	The lesion completely disappeared
Kim et al[4]	F/7 months	5 months of age	Dorsal side of the right little finger over the DIP joint	Partial invasion toward the nail plate	None	The lesion remained
Hughes et al[6]	F/23 months	At birth	Radial aspect of right third finger over the DIP joint	Extend to collagen ligament	Excisional biopsy	No recurrence
Esterly et al[7]	M/2 years	3 months of age	Base of left little finger	Extensive facial eruption	None	No F/U
Chang et al[8]	M/2.5 years	Unknown	Right index finger beneath a finger	Lifting up and dystrophic nail by tumor	Tumor removal after nail avulsion	No F/U

cytic proliferation[3].

Only six cases of JXG of the fingers have been reported in the English literature; these cases are summarized in Table 1[1,3,4,6-8]. Sonoda et al[1] reviewed 57 patients with JXG, and reported the case of a 27-year-old woman with JXG involving a finger. Piraccini et al[3] presented a patient with JXG on the proximal nail fold of the right thumbnail. Kim et al[4] and Hughes et al[6] each encountered a case of JXG mimicking a giant cell tumor of the tendon sheath (GCTTS), which had extended to the collateral ligaments. Esterly et al[7] described a 2-year-old boy with an extensive facial eruption. Initially, two biopsies were taken from the right cheek, and pathology results were interpreted as being compatible with histiocytosis X. However, the clinical course, nature of the eruption, and laboratory findings all argued against a diagnosis of histiocytosis X. Upon further evaluation, a reddish papule was detected at the base of the left fifth finger, which was histologically diagnosed as JXG. Another report by Chang et al[8] documented a 2.5-year-old Caucasian male with JXG in the nail bed beneath a fingernail, which was presented with progressive dystrophy and was elevated by the tumor in the nail bed.

A histological study of JXG revealed an ill-defined, unencapsulated, dense histiocytic infiltrate in the papillary and reticular dermis[2]. Neutrophils, eosinophils and lymphocytes were observed scattered within the lesion[2]. Mature lesions were reported to contain Touton giant cells, foreign body giant cells, and foamy cells[6]. Touton giant cells are characterized by a peripheral rim of vacuolated cytoplasm surrounding a ring of nuclei bordering a central zone of eosinophilic cytoplasm, a feature that is nearly pathognomonic for JXG[6]. Serum lipid profiles are usually normal and laboratory evaluations are not mandatory[6]. Radiologically, JXG does not involvement of the underlying bony structure[6].

JXG on the finger is often misdiagnosed clinically as a giant cell tumor of the tendon sheath, dermatofibroma, or as infantile digital fibroma[4,6,9]. However, these diagnoses can be distinguished from JXG by the absence of Touton giant cells, which are the histological hallmark of JXG[9]. Notably, the clinical patterns of JXG and solitary reticulohistiocytoma are similar[4]. Solitary reticulohistiocytoma is a rare, benign disorder of the non-Langerhans cell histiocytic family, like JXG. Histologically, histiocytes form an abundant, smooth, eosinophilic "ground-glass" cytoplasm in solitary reticulohistiocytoma[2,9]. However, the histological findings of the lesion in our patient revealed few giant cells with ground glass cytoplasm, and therefore, we diagnosed JXG.

The cutaneous lesions of JXG regress spontaneously within 3 to 6 years[6]. However, hyperpigmentation, atrophy, or anetoderma may remain in up to 48% of cases after regression[6]. Nevertheless, despite the self-limited nature of JXG, surgical intervention is usually considered for cosmetic or diagnostic reasons[4]. For our patient, the lesion was totally removed with a 4 mm punch for diagnostic purposes.

In conclusion, the clinical diagnosis of JXG is easily made. However, JXG can develop in unusual sites with equally unusual shapes, distributions, or sizes[6]. JXG should be considered during the differential diagnosis of a soft tissue tumor of the finger in children[4]. If physical and radiology examinations do not suggest a diagnosis, a histopathological examination is mandatory. This case illustrates a rare JXG of the right fourth finger in a 4-year-old girl, which was diagnosed based on its characteristic histological findings.

REFERENCES

1. Sonoda T, Hashimoto H, Enjoji M. Juvenile xanthogranuloma. Clinicopathologic analysis and immunohistochemical study of 57 patients. Cancer 1985;56:2280-2286.
2. Newman B, Hu W, Nigro K, Gilliam AC. Aggressive histiocytic disorders that can involve the skin. J Am Acad Dermatol 2007;56:302-316.
3. Piraccini BM, Fanti PA, Iorizzo M, Tosti A. Juvenile xanthogranuloma of the proximal nail fold. Pediatr Dermatol 2003;20:307-308.
4. Kim EJ, Kim MY, Kim HO, Park YM. Juvenile xanthogranuloma of the finger: an unusual localization. J Dermatol 2007;34:590-592.
5. Hernandez-Martin A, Baselga E, Drolet BA, Esterly NB. Juvenile xanthogranuloma. J Am Acad Dermatol 1997;36:355-367.
6. Hughes DB, Hanasono MM, Nolan WB 3rd. Juvenile xanthogranuloma of the finger. Pediatr Dermatol 2006;23:53-55.
7. Esterly NB, Sahihi T, Medenica M. Juvenile xanthogranuloma. An atypical case with study of

ultrastructure. Arch Dermatol 1972;105:99-102.
8. Chang P, Baran R, Villanueva C, Samayoa M, Perrin C. Juvenile xanthogranuloma beneath a fingernail. Cutis 1996;58:173-174.
9. Elder DE, Elenitsas R, Johnson BL Jr, Murphy GF. Lever's histopathology of the skin. 9th ed. Philadelphia: Lippincott Williams & Wilkins, 2005: 686-688.