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.

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. The pathophysiology of JXG is not well understood, although it is thought to originate from a histiocytic granulomatous reaction.

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

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
An Unusual Juvenile Xanthogranuloma on a Finger 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-

---

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

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

---

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).
cytic proliferation.

Only six cases of JXG of the fingers have been reported in the English literature; these cases are summarized in Table 1. Sonoda et al reviewed 57 patients with JXG, and reported the case of a 27-year-old woman with JXG involving a finger. Piraccini et al presented a patient with JXG on the proximal nail fold of the right thumbnail. Kim et al and Hughes et al 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 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 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. Neutrophils, eosinophils and lymphocytes were observed scattered within the lesion. Mature lesions were reported to contain Touton giant cells, foreign body giant cells, and foamy cells. 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. Serum lipid profiles are usually normal and laboratory evaluations are not mandatory. Radiologically, JXG does not involve ment of the underlying bony structure.

JXG on the finger is often misdiagnosed clinically as a giant cell tumor of the tendon sheath, dermatofibroma, or as infantile digital fibroma. However, these diagnoses can be distinguished from JXG by the absence of Touton giant cells, which are the histological hallmark of JXG. Notably, the clinical patterns of JXG and solitary reticulohistiocytoma are similar. 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. 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. However, hyperpigmentation, atrophy, or anetoderma may remain in up to 48% of cases after regression. Nevertheless, despite the self-limited nature of JXG, surgical intervention is usually considered for cosmetic or diagnostic reasons. 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. JXG should be considered during the differential diagnosis of a soft tissue tumor of the finger in children. 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

7. Esterly NB, Sahib T, Medenica M. Juvenile xanthogranuloma. An atypical case with study of