A Case of Unusual Juvenile Xanthogranuloma on the Subungual Area

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Dear Editor:

Juvenile xanthogranuloma (JXG) is a benign histiocytic proliferative disorder of skin, which is characterized by solitary or multiple yellow-red nodules. JXG usually occurs in infancy or early childhood. The pathophysiology of JXG is not yet understood, although it is thought to repre-

sent a histiocytic granulomatous reaction. Lesions mostly present on the head, neck, and trunk, however, the involvement of the distal extremities such as fingers and toes is very rare; only 6 cases of JXG involving a nail have been reported in the English and Korean literature. Herein, we report on an unusual clinical presentation of JXG de-

Fig. 1. (A) Yellow firm nodule under the nail right 3rd finger nail plate. (B) Dense intradermal histiocytic infiltrates and a number of giant cells (H&E, $\times 200$). (C) Foreign body giant cells with haphazard nuclear arrangement were mainly observed (H&E, $\times 400$). (D) All of histiocytes were positive for CD163 (CD163, $\times 400$).

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veloping below the finger nail.

JXG is a benign histiocytic proliferative disorder of skin, which is characterized by solitary or multiple yellow-red nodules. JXG usually occurs in infancy or early childhood. Lesions mostly present on the head, neck, and trunk; however, the involvement of the distal extremities such as fingers and toes is very rare; only 6 cases of JXG involving a nail have been reported in the English and Korean literature¹⁻⁵. Herein, we report on an unusual clinical presentation of JXG developing below the finger nail.

A healthy 7-month-old girl presented with 4 months history of a yellow discolored nail deformity in the right 3rd finger. There was no remarkable past or family history. On physical examination, the yellow firm nodule was seen under the nail plate and a yellowish plaque was also observed on the neck (Fig. 1A). We received the patient's consent form about publishing all photographic materials. There was a yellowish patch on the neck on physical examination. Excisional biopsy was done to rule out viral wart or corn.

Pathologic finding reveal showed dense intradermal histiocytic infiltrates which contained foamy cell (Fig. 1B). Many giant cells were observed, which mainly comprised of foreign body giant cells with haphazard nuclear arrangement and focal Langhans type giant cells with horse-shoe-shape nuclear arrangement (Fig. 1C). Differential diagnosis included non-Langerhans cell histiocytosis such as JXG and soft tissue mesenchymal tumors with rich giant cells such as giant cell tumor of soft tissue. All histiocytes

was positive for CD163 (Fig. 1D). There were scattered lymphocytes and eosinophils and were not chondrocytes and osteocytes. Based on histopathologic findings, age and morphology, it was diagnosed with JXG. When she came back 15 months after the skin biopsy, there was no residual tumor.

JXG is the most common form of non-Langerhans cell histiocytosis and is considered a benign histiocytic proliferation. JXG usually present on the head, neck, and trunk, however, the involvement of fingers and toes is rare and few cases were reported. Only 6 cases of JXG involving nail unit have reported in the English and Korean literature; these cases are summarized in Table 1¹⁻⁵.

Four cases²⁻⁵ were associated with a finger nail and two cases^{1,5} involved a toe nail. Two cases^{4,5} involved proximal nail fold and other cases^{1-3,5} occurred in subungual area. JXG regressed spontaneously in two cases^{2,5}. Tumor removal was done in three cases^{1,2,5}; permanent nail dystrophy was observed in one case¹ and no recurrence was observed in another case⁵.

JXG associated with nail unit is often misdiagnosed clinically as a common wart, glomus tumor, pyogenic granuloma, keratoacanthoma, or as corn⁵. However, these diagnoses can be distinguished from JXG by the characteristic histopathologic findings; dense intradermal histiocytic infiltrates which contained foamy cells and giant cells. In some cases, immunohistochemistry such as factor XIIIa and CD163 could be helpful in differentiating other fibrohistiocytic tumors.

Table 1. Review of the cases of juvenile xanthogranuloma involving the nail reported in the English and Korean language published work

Case	Sex/age	Onset	Location	Associated features	Treatment	Outcome	Reference no.
1	M/3 yr	Before 1.5 years	Left second toe beneath a toenail	The whole nail bed including the matrix was involved	Tumor removal	Permanent nail dystrophy after 7 months	1
2	M/2.5 yr	Unknown	Right index finger beneath a fingernail	Lifting up and dystrophic nail by tumor	Tumor removal after nail avulsion	No F/U	2
3	M/18 mo	Before 7 months	Proximal nail fold of right thumb nail	Severe depressed nail plate and hyperkeratotic cuticle	None	The lesion completely disappeared	3
4	F/7 mo	Before 5 months	Proximal nail fold of right little finger	Partial invasion toward the nail plate	None	The lesion remain 5-month- F/U	4
5	F/24 mo	Before 1 month	Left First toe beneath a toenail	The whole nail bed including the matrix was involved and nail lost	None	The lesion completely disappeared	5
6	F/21 yr	Before 6 months	Left thumb finger beneath a umb nail	Lifting up by tumor	Tumor removal after nail avulsion	No recurrence	5
7	F/7 mo	Before 4 months	Right third finger beneath a fingernail	Lifting up and dystrophic nail by tumor	Tumor removal without nail avulsion	No recurrence	This case

M: male, F: female, F/U: follow-up.

In conclusion, JXG can present in unusual sites associated with nail unit and can be misdiagnosed such as common wart and other diseases. So, it is necessary to check histopathology not to misdiagnose and mistreat. This case illustrates a rare JXG of the right 3rd finger nail bed in a 7-month-old girl which was diagnosed based on its characteristic histopathologic findings.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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