QUIZ SECTION

Dystrophy and Yellowish Discolouration of a Finger-nail in a 22-month-old Infant: A Quiz

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A 22-month-old female infant presented with distal dystrophy of the nail plate of her right index finger, which had persisted for 5 months. The nail also showed longitudinal ridging, onycholysis, and a small yellow spot on the lunula (Fig. 1A). She did not have any underlying diseases. No symptoms and signs, including itching, oozing, pain, and tenderness, were reported. Dermoscopy showed the presence of a relatively well-defined homogenous, bright-yellow circular area in the proximal area of the nail plate (Fig. 1B). Ultrasound revealed a 4.4×1.9×3.4-mm soft-tissue mass with calcified spots between the nail and the distal phalangeal bone. However, no specific bony abnormalities were identified on finger X-ray, and no abnormal laboratory results associated with calcium metabolism were obtained.

What is your diagnosis? See next page for the answer.

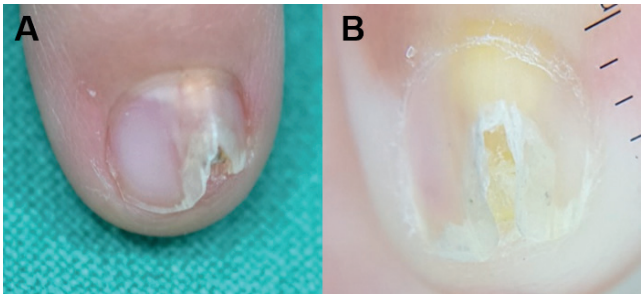

Fig. 1. (A) An asymptomatic solitary yellow spot on the lunula and destruction of the distal nail plate of the right index finger. (B) Dermoscopic findings of the nail unit showing a relatively well-defined, homogenous, bright-yellow circular area, onycholysis, and distal longitudinal nail destruction.

ANSWERS TO QUIZ

Dystrophy and Yellowish Discolouration of a Finger-nail in a 22-month-old Infant: A Commentary

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Diagnosis: Subungual juvenile xanthogranuloma

Excisional biopsy was performed to obtain an accurate diagnosis and to repair the nail deformity. Under general anaesthesia, her nail was lifted up using the window technique, revealing a well-defined, round, yellow mass (Fig. 2A). The mass was located above the distal nail matrix, and was completely removed. Histological analysis of the excised tissue showed dense lymphohistiocytic proliferation and some Touton giant cells and foamy histiocytes with vacuoles in the tumour (Fig. 2B). On immunohistochemical staining, the tumour was found to be positive for CD68 (Fig. 2C), but negative for S-100 and cytokeratin. A diagnosis of subungual juvenile xanthogranuloma (JXG) was made. Fourteen months after the surgery, a distally elongated lunula was observed with a small triangular shape, without recurrence or sequelae, including nail deformity or onycholysis (Fig. 2D).

Various subungual tumours that can cause nail deformities in children include epidermal cyst, subungual exostosis, haemangioma, and pyogenic granuloma. An epidermal cyst rarely involves the subungual area, but is almost always accompanied by a history of trauma. A subungual exostosis mainly involves the great toenails. Not only the bone, but also the bony tumour, is visible on X-ray. A subungual haemangioma can involve any soft tissue, but shows a purple-reddish colour change, which disappears when compression is applied. A pyogenic granuloma is a solitary mass with a bright-red colour, which bleeds easily.

This case was distinguished from all of the above-mentioned conditions. Nail deformity was noted, but it was an asymptomatic and proximally located yellow subungual mass in a young child. The tumour was diagnosed as JXG on histological examination. However, the dermoscopic findings did not correspond to JXGs in other locations, which have a red-yellow centre, discrete erythematous halo, or branched telangiectasia (1). The authors hypothesized that the tumour was compressed by the nail, and the vascular component disappeared on dermoscopic evaluation.

JXG is a type of non-Langerhans cell histiocytosis. It usually presents on the face, neck, and upper trunk, but rarely on the nail units (2, 3). JXG lesions undergo spontaneous involution over a period of 3–6 years, but if any cosmetic concern or risk to vital functions exists, the lesion should be treated (4). The treatment of choice of cutaneous JXG is surgical excision, and CO_2 laser, intralesional steroid injection, cryotherapy, or low-dose radiotherapy are treatment alternatives (3). However, the subungual area is a rare location for JXG. Furthermore, accurate diagnosis is sometimes difficult because of the subungual location of the tumour and due to concerns about cosmetic disfigurement, i.e. the potential for the nail deformity to become more severe;

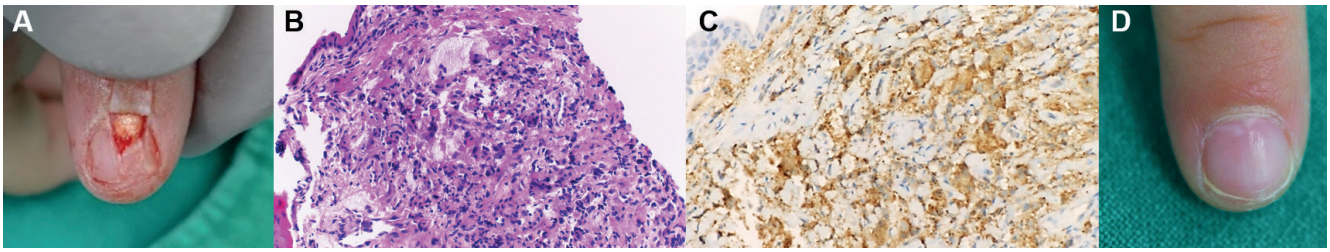

Fig. 2. (A) After lifting up the nail, a well-circumscribed, round, yellow mass on the distal nail matrix was noted. (B) Lymphohistiocytic proliferation with some Touton giant cells and foamy histiocytes with vacuoles in the tumour (haematoxylin and eosin (H&E), 100×). (C) The tumour was positive on CD68 immunohistochemical staining (200×). (D) On the 14-month follow-up after excision, no recurrence and no nail deformities were found. Distally elongated lunula was observed with a small triangular shape.

Table I. Cases in the literature of juvenile xanthogranuloma involving the nail unit

Case	Sex/age	Duration	Location	Appearances and nail deformity	Treatment	Follow-up	Ref.
1	Male/3 years	1.5 years	Left second toenail	Tumour involving the whole nail bed with nail deformity	Tumour removal	No recurrence Permanent nail dystrophy	5
2	Male/2.5 years	Unknown	Right index fingernail	Yellowish hue on the nail plate with dystrophic nail lifted up by tumour	Tumour removal after nail avulsion	No follow-up	6
3	Male/18 months	7 months	Proximal nail fold of right thumb	Pink-red tumour involving the whole proximal nail fold with severely depressed nail plate	None	Spontaneous remission after 31 months without complication	7
4	Female/7 months	5 months	Proximal nail fold of right little finger	Reddish, dome-shaped tumour involving the nail plate causing nail deformity	None	Persisting during the 5 months of follow-up period	8
5	Female/24 months	1 month	Left great toenail	Yellowish to erythematous papule on the whole nail bed with loss of nail plate	Partial removal during biopsy	Spontaneous remission after 16 months without complication	9
6	Female/21 years	6 months	Left thumbnail	Yellow spot on the nail bed with longitudinal ridging and onycholysis	Tumour removal after nail avulsion	No recurrence or complication	9
7	Female/7 months	4 months	Right third fingernail	Yellow firm nodule on the distal nail bed with distal nail plate destruction	Tumour removal without nail avulsion	No recurrence or complication	10
8	Female/22 months	5 months	Right index fingernail	Yellow spot on the nail lunula with distal nail plate destruction, longitudinal ridging, and onycholysis	Tumour removal with window technique	No recurrence or complication	Current case

subungual JXGs often need surgical excision.

Including this case, only 8 cases (5–10) of JXG involving the nail unit have been reported, as summarized in Table I. These cases mainly involved children under 3 years of age (3 males and 5 females). Nail deformity was observed in all patients, and yellow lesions were noted in 5 of the 8 cases. All patients visited a hospital and received a diagnosis within 7 months. The timely hospital visit in each case was a result of changes in the appearance of the nail unit area due to the tumours. Three cases of subungual JXGs were not treated, whereas tumour removal was performed in the other 5. Of the 3 cases in which the tumour was not treated, spontaneous remission was observed in 2 cases, at 16 and 31 months. Of the 5 cases in which the tumour was removed, 4 cases did not recur, whereas the fifth case was lost to follow-up. Permanent nail dystrophy after tumour removal was observed only in 1 out of the 5 cases.

In conclusion, even if dermoscopic findings do not correspond to known JXG presentations, if a nail deformity with a yellow discolouration of the nail plate is observed in a young child, subungual JXG should be considered as a differential diagnosis, and a surgical approach would provide not only an accurate diagnosis but also a correction of the nail deformity.

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The authors have no conflicts of interest to declare.

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