Periungual squamous cell carcinoma in an 8-year-old Hispanic boy

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INTRODUCTION

Squamous cell carcinoma (SCC) of the nail unit is a relatively rare diagnosis that mostly affects men in the fifth and sixth decades of life.1,2 Tumors of the nail unit often begin in the lateral nail grooves or folds and are frequently misdiagnosed as more common dermatoses such as dermatitis, viral warts, or fungal infection; therefore, late diagnosis (average 5–7 years) is commonplace.1-3 It is usually not until tumor spread or numerous treatment failures that a correct diagnosis is made via adequate biopsy and histopathologic evaluation.3

Given the variety of presentations and low prevalence of nail SCC, especially in the pediatric population, identifying common diagnostic features is imperative so that early diagnosis may be achieved. Risk factors for SCC development include ionizing radiation, sunlight, human papillomavirus (HPV) infection (often high-risk types), arsenic exposure, trauma, chronic paronychia, and immunosuppression.1,2

We report a case of periungual invasive squamous cell carcinoma in an 8-year-old Hispanic boy. To date, this is the youngest case reported in the literature to our knowledge.

REPORT OF A CASE

An 8-year-old Hispanic boy presented to the pediatric dermatology clinic for evaluation of dark pigmentation of his fingernail. Per the patient's mother, the patient began to manipulate the cuticle on the second digit of the right hand several months before presentation. A dark bump was then noted as well as linear discoloration of the nail plate. The patient denied any discomfort or bleeding at the site. There was no family or personal history of warts or skin cancer or personal history of immunosuppression. The patient had not received the HPV vaccine, and the lesion was not previously treated as a wart.

On examination of the right index finger, a black-brown papule was seen overlying the lateral proximal nail fold, which showed superficial scaling, lateral longitudinal melanonychia, and ridging of the nail plate (Fig 1). Because of the patient's age, biopsy was performed in the operating room under general anesthesia. A lateral longitudinal nail unit excision was performed. Histologic examination found an atypical proliferation of squamous cells with scattered and marked nuclear pleomorphism, frequent mitotic figures, and dyskeratotic cells (Fig 2). Cytologic features suggestive of HPV were seen. HPV genotyping by polymerase chain reaction was positive for a type other than those represented in the group reported (types 6, 11, 16, 18, 31, 33, 35, 39, 45).

Once the diagnosis of periungual SCC was made, appropriate definitive management was achieved utilizing Mohs micrographic surgery under local anesthesia in an outpatient setting. Distraction techniques and topical anesthetic were necessary to keep the patient comfortable. The tumor was

ABBREVIATIONS USED

HPV: human papillomavirus
SCC: squamous cell carcinoma
removed successfully in 2 stages and healed by secondary intention without incident (Fig 3). Three years after the procedure, there is no evidence of recurrence.

**DISCUSSION**

The typical patient with SCC of the nail unit is a male in the fifth or sixth decade of life, with a thumb or index fingernail affected. Definitive diagnosis is often delayed for years and can result in more advanced disease presentation. Our case represents a unique outlier from the typical population given our patient’s young age and relatively short time to diagnosis. Based on our review of the literature, this is the fifth case of a pediatric nail unit SCC and the youngest invasive periungual SCC case to date (Table 1).4-8 Presenting signs of these pediatric cases include a growing brown scaly plaque at the lateral nail fold, a pseudofibrokeratoma of the subungual area of the distal nail plate, and recurrent paronychia, respectively. In our case, a scaly hyperpigmented papule at the base of the nail plate and longitudinal melanonychia and nail dystrophy were the presenting signs. The pigmentation and nail dystrophy were concerning and led to rapid biopsy of the lesion. Oncogenic strains of HPV have been implicated in the pathogenesis of nail SCC. In a case series of 103 adults (ages 22–89), HPV16 was implicated 74% of the time.2 In addition, 33% of the tumors presented as periungual verrucal masses. A separate case series, including SCCs from all areas of the hand, showed that all 6 periungual SCCs examined were caused by highly oncogenic strains of HPV.9 Only one of the other 19 nonperiungual SCCs in the study were caused by highly oncogenic strains of HPV, indicating a unique immunologic susceptibility of the periungual region of the hand.9 Our case of periungual SCC was likely HPV induced, but the specific subtype was not identified or typical of known oncogenic subtypes. The patient’s health plan refused to approve further HPV subtyping.

Our patient provided a unique challenge because of his young age and lack of treatment.
guidelines of nail SCC in this age group; maintaining full utilization of the digit was imperative, and we wished to avoid radical surgery. Local and wide excision, Mohs micrographic surgery, distal digital amputation, and topical chemotherapy (for in situ disease) are all described in the treatment of nail SCC. When bony involvement occurs, partial digital amputation is warranted. Mohs micrographic surgery is the most effective technique at maintaining function and preventing recurrence. A meta-analysis that evaluated more than 100 cases of HPV-induced digital SCC (57 cases reported as subungual/periungual) found that Mohs micrographic surgery was the most effective treatment but with a 20% recurrence rate.

Given the variable presentation and frequent misdiagnosis of nail unit SCC, our case shows the importance of high clinical suspicion and early biopsy for clinically atypical periungual lesions in the pediatric population. We also describe the use of Mohs micrographic surgery in an 8-year-old boy to successfully treat invasive periungual SCC.

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Table I. Literature review of pediatric nail unit squamous cell carcinoma

| Case | Sex/Age (y) | Location | Presentation | HPV Treatment/Result |
|------|------------|----------|--------------|----------------------|
| Wöckel et al, 1979 | Female/10 | Nail bed, right thumb | Recurrent paronychia | Not stated |
| Dominguez-Cherit et al, 2003 | Female/13 | Subungual, left index finger | Pseudo-fibrokeratoma of the distal nail bed | Negative (tested types not specified) |
| Kim et al, 2013 | Female/4 | Periungual, right lateral finger | 5-mm brown macule extending under the nail plate (SCC in situ) | Negative for types 6, 11, 16, 18, 21, 33, and 55 |
| Hyun et al, 2016 | Male/12 | Periungual, right index finger | Brown plaque adjacent to the proximal nail fold (SCC in situ) | Positive for unknown type, negative for types 6, 11, 16, 18, 31, 33, 35, 45 |
| Kelling et al, 2016 (Current patient) | Male/8 | Periungual, right index finger | Scalpel hyperplastic papule, longitudinal melanonychia, nail dystrophy | Positive for unknown type, negative for types 6, 11, 16, 18, 31, 33, 35, 45 |

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