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Case Report

A case report of onycholemmal carcinoma in a 61-year-old Chinese male

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ABSTRACT

Onycholemmal carcinoma is characterized as a slowly progressing malignant tumor originating from the epithelium of the nail bed. A limited number of cases have been documented in the English literature. The majority of the patients included in the reports underwent amputation of the affected phalanx, and no instances of recurrence were noted during the follow-up period. A 61-year-old Chinese male presented with a persistent ulceration on the nail bed of the right great toe. Microscopic analysis indicated the presence of an epithelial tumor consisting of small keratocysts with sudden central keratinization and atypical keratinocyte nests that were devoid of a granular layer. The tumor exhibited infiltrative growth within the dermis, displaying a multilobulated pattern, but did not extend into the distal phalangeal bone. Based on these findings, the diagnosis of onycholemmal carcinoma was made for this case. All documented cases indicate that onycholemmal carcinoma is a rare malignant tumor originating from the nail bed epithelium, and its clinical progression is typically slow and non-aggressive. This case is presented to provide an analysis of the clinical and pathological features of onycholemmal carcinoma, aiming to assist in the clinical selection of treatment options.

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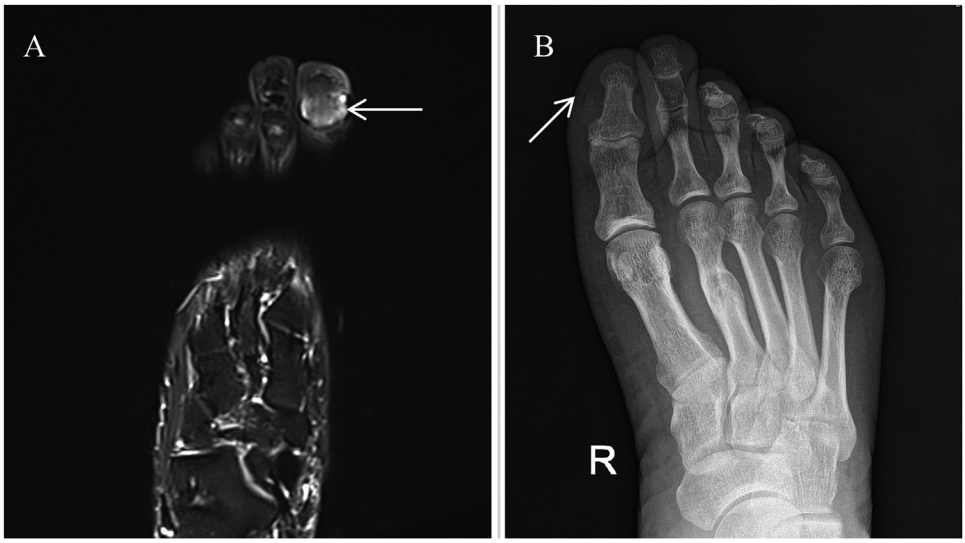

Figure 1. Coronal fat-suppressed T2WI showing a long flaky T2 signal (arrow) at the distal nail bed of the right great toe (A). The X-rays showing a slightly rough bone density in the distal phalanx of the right great toe, along with swollen soft tissues around the distal phalan and punctate cacification (arrow) (B).

Case report

In June 2020, a male patient aged 61 years was admitted to our hospital with a persistent ulceration of the nail bed on the right great toe. The ulceration was painless and had been gradually increasing in size over the previous two years. The patient had previously undergone a histopathologic biopsy at another hospital before being admitted to our facility. The biopsy results indicated the presence of moderate to severe atypical hyperplasia in squamous cells, with the initial diagnosis being squamous cell carcinoma. Regrettably, there were no pathological sections available. The biochemical parameters of the blood remained within the normal range. The patient did not have a history of injury or infection.

The MRI conducted in the hospital revealed an anomalous signal in the distal nail bed of the right great toe, measuring 1.8 × 1.4 cm. Coronal fat-suppressed T2-weighted imaging revealed a long, flaky T2 signal (Figure 1A) at the distal nail bed of the right great toe, with a clearly defined boundary. The integrity of the bone structure was preserved, and the signal exhibited uniformity throughout the bones. The X-rays revealed a slightly rough bone density in the distal phalanx of the right great toe, along with swollen soft tissues around the distal phalanx and punctate calcification (Figure 1B)

Consequently, the patient underwent the amputation of the affected distal phalanx. The gross examination revealed that the resected specimen measured 3.5 × 3.0 × 1.0 cm in size, and exhibited an ulcer measuring 2.1 × 1.8 cm on the surface of the nail bed (Figure 2). Microscopic analysis demonstrated an infiltrative proliferation of epithelial tumor, consisting of atypical keratinocyte nests without a granular layer and small keratocysts with sudden central keratinization (Figure 3). The tumor cells may have originated from the epithelium of the superficial nail bed and exhibit onycholemmal characteristics. The tumor exhibited infiltrative growth within the dermis, displaying a multilobulated pattern, but did not extend into the distal phalangeal bone. The tumor did not involve the excision margins. The patient was monitored for a period of two years, during which no instances of local recurrence or inguinal lymph node metastasis were detected.

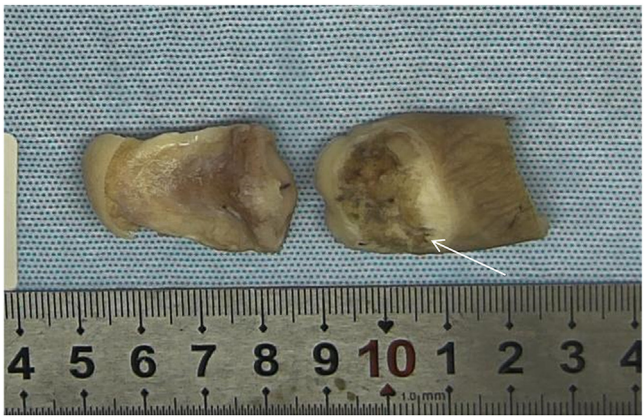

Figure 2. The gross of the excised specimen indicated the presence of an ulcer on the nail bed surface (arrow).

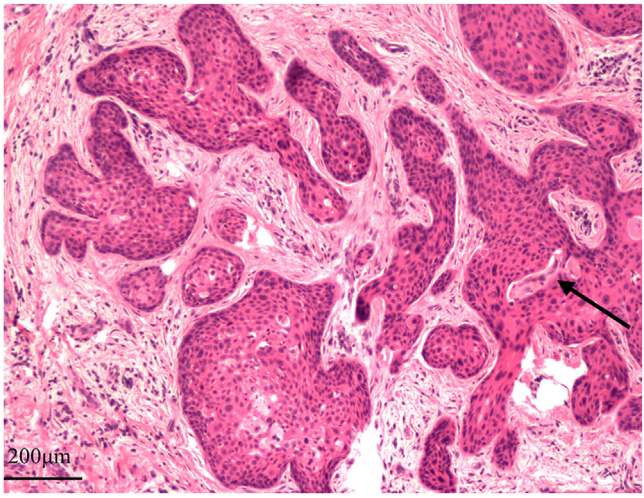

Figure 3. The infiltrative growth of atypical keratinocyte nests without a granular layer and a small keratocyst (arrow) with sudden central keratinization lined by atypical keratinocytes.

Discussion

Onycholemmal carcinoma is an infrequent malignant neoplasm originating from the epithelium of the nail bed. Fewer than nine cases are currently documented in the English literature.[1–8] The documented instances primarily affected elderly individuals, and the disease's progression was characterized by slow development of distal digit lesions accompanied by nail bed ulceration, paronychia, onycholysis, mild pain, and swelling.[1-3,5,6] Alessi et al. (year) discovered that the nail bed exhibited similarities to the outer root sheath of the hair follicle. They identified a malignant tumor originating from the nail bed epithelium and named it "onycholemmal carcinoma".[1]

Histopathologically, onycholemmal is characterized by showed infiltrative infiltrative growth of atypical keratinocyte that lack a lacking layer, as well as and small keratocysts with abrupt central keratinization. Due to the challenges in differentiating the microscopic characteristics of onycholemmal carcinoma from squamous cell carcinoma, some researchers have categorized onycholemmal carcinoma as a rare subtype of squamous cell carcinoma.[9] When the keratocyst exhibits prominent fea-

tures, the tumor can be classified as a malignant proliferating onycholemmal cyst.[10] When diagnosing onycholemmal carcinoma, it is important to exclude squamous cell carcinoma, basal cell carcinoma, Bowen disease, and subungual keratoacanthoma.[2,6] Maffeis et al. (year) conducted molecular profiling of a reported case and identified potential associations between onycholemmal carcinoma pathogenesis and alterations in the p16/RB/E2F pathway.[8]

At present, a no standard treatment for onycholemmal carcinoma has not been established. The majority of the patients included in the reports underwent amputation of the affected phalanx, and no instances of recurrence were observed during the follow-up period. Due to the indolent natural clinical course of the tumor, some scholars advocate non-amputation treatments in order to maintain the function of the patient's digits. Non-amputation treatments, such as radiation and Mohs micrographic surgery, have been documented in the literature.[5] Upon reviewing our case, it appears that the treatment options for amputation may have been slightly excessive. The preoperative MRI and X-rays images of the patient showed that the lesion was located in the dermis, did not extend into the distal phalangeal bone, and there was no local lymph node metastasis. Clinicians can consider radiation therapy or Mohs micrographic surgery, which are less invasive, toe-preserving treatment options to help the patient reduce functional loss. Our case also suggests that a clear biopsy diagnosis and the clinician's understanding of the clinicopathological features of the tumor before treatment are helpful for developing a rational treatment plan.

Conclusions

Onycholemmal carcinoma is an uncommon malignant tumor originating from the epithelium of the nail bed, and it typically follows an indolent clinical course. Currently, there is a deficiency in standardized clinical treatment for onycholemmal carcinoma. It is also necessary to gather additional cases of onycholemmal carcinoma, compile its clinical and pathological characteristics, and conduct further research and discussion on the established findings.

Declaration of competing interest

The authors declare there is no conflict of interests.

Informed consent

Informed consent was obtained from the patient in this study.

Ethical approval

This study was approved by Hefei BOE Hospital Ethics Committee.

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None.

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