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Koenen Tumors Predisposing to Recurrent Onychomycosis: An Interesting Case

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Case Report
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Abstract

Koenen tumors are uncommon nail tumors seen characteristically in patients of tuberous sclerosis. They cause longitudinal nail grooves, onycholysis, nail deformity or nail dystrophy which may predispose the affected nails to secondary infections. We report a case of a 35-year-old male with multiple koenen tumours having recurrent onychomycosis. A 35-year-old male presented with whitish discoloration and loss of distal part of nails from 2-3 years and scaling on right hand and feet from 1 year. On examination, there was scaling over dorsal aspect of right hand and both feet. There was subungual hyperkeratosis, distal onycholysis and distal nail dystrophy and nail plate splitting in multiple nails. Digitate growths arising from proximal nail fold (Koenen tumours) were also noted. KOH examination from skin and nails demonstrated fungal hyphae. Patient recounts taking multiple courses of topical and oral antifungals giving temporary relief but recurrence of lesions occurs after stopping treatment. It is possible that Koenen tumours themselves or the nail changes caused by them like nail plate thinning, depression or dystrophy act as portal of entry for fungus and thus, predispose to recalcitrant, chronic or recurrent fungal nail infections. Similar cases of onychomycosis coexisting with other nail tumors have also been reported. There is a need of more studies on association of nail tumors with onychomycosis as a risk factor and management in these cases.

Keywords: Nails; Onychomycosis; Tumors

Introduction

Koenen tumors are benign, slow growing tumors of the nail unit included among the diagnostic criteria of tuberous sclerosis. Depending on the location of the tumor, they may lead to nail plate thinning, depression in the nail plate or even nail dystrophy; thus possibly predisposing to invasion by pathogens. We report a case of a 35-year-old male with multiple koenen tumours and recurrent onychomycosis.

Case Report

A 35-year-old male presented with whitish discoloration and loss of distal part of nails from 2-3 years and scaling over both his hands and feet from 1 year (Figure 1A). There was history of splitting and deformity in lateral part of nails from 10 years for which patient did not take any treatment as the lesions were asymptomatic. On examination, there was scaling over dorsal aspect of right hand and both feet. On nail examination, subungual hyperkeratosis, distal onycholysis, distal nail dystrophy and nail plate splitting in multiple nails were observed in Figure 1B. On onychoscopy, digitate growths arising from proximal nail fold in addition to the above findings were noted Figures 2A & 2B. Patient also had multiple papules on face suggestive of angiofibromas and was under follow up from Neurology for seizures (Figure 3). KOH mount of scrapings from skin and nail demonstrated fungal hyphae. A diagnosis of Koenen tumors

with coexistent onychomycosis (tinea unguum) and tinea pedis/mannum was made. Patient recounts taking multiple courses of topical and oral antifungals for nails in last 2-3 years giving temporary relief but recurrence of lesions after stopping treatment. From 1 year, patient developed scaling first on hand and then on feet. There was no other identifiable risk factor for recurrent onychomycosis. Patient was started on itraconazole 200 mg. Shave excision followed by phenolization was planned for Koenen tumors.

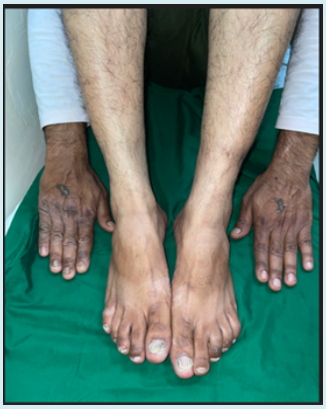

Figure 1A: Scaling over hands and feet.

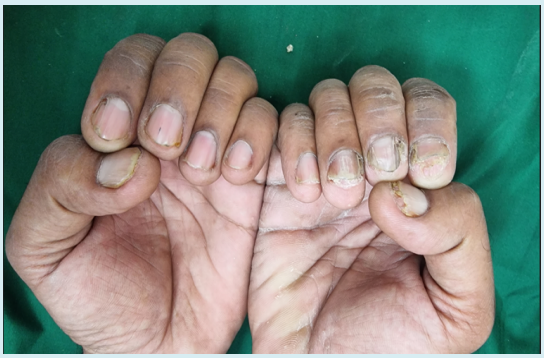

Figure 1B: Subungual hyperkeratosis, distal onycholysis, distal nail dystrophy and nail plate splitting in multiple nails.

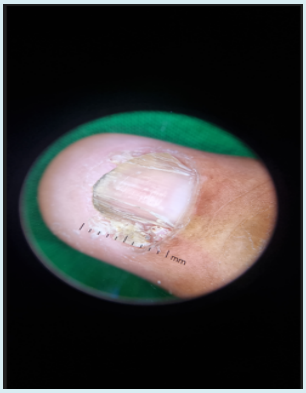

Figure 2A: Flesh coloured excrescences arising from proximal nail fold with nail plate dystrophy.

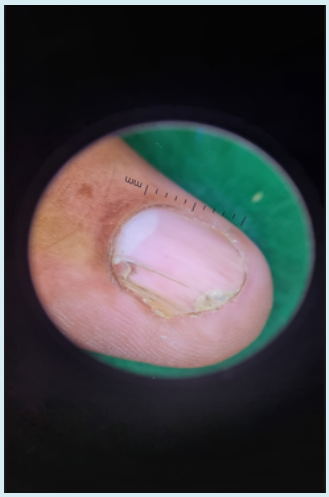

Figure 2B: Small outgrowth seen in proximal nail fold with longitudinal grooves and splits.

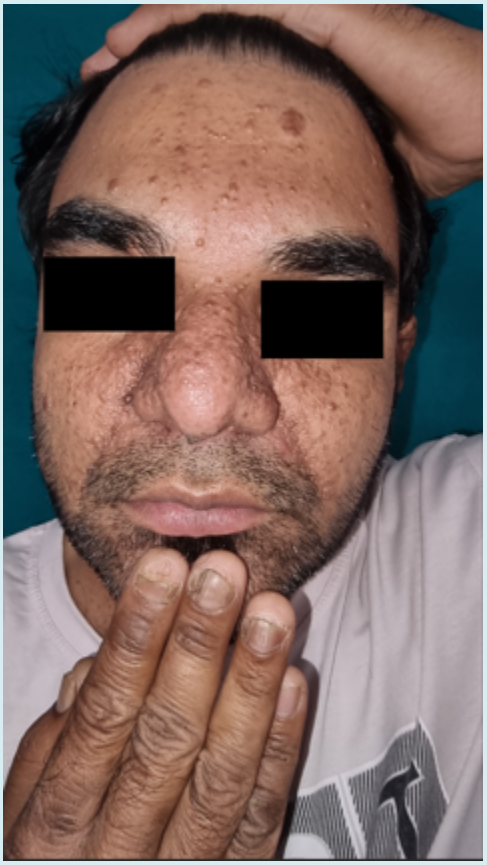

Figure 3: Facial angiofibromas and koenen tumours in a patient of Tuberous Sclerosis.

Periungual/subungual fibromas or Koenen tumors are pathognomic manifestation of tuberous sclerosis, an autosomal dominant genetic disorder. They are more

common in females, arise around puberty and affect toes more than fingers [1]. Clinically, they present as skin-coloured or reddish nodules arising from the periungual or subungual part of lateral nail groove, nail plate, or proximal nail fold [2]. Some other nail findings include longitudinal nail grooves, longitudinal erythronychia and longitudinal leukonychia. The lesions often become keratotic and grow in size to cause onycholysis, nail deformity or even complete nail dystrophy. Onychoscopy makes it easier to identify the flesh coloured excrescences and longitudinal striations which may be subtle in early cases. Typical garlic clove morphology has been also described in few cases consisting of a yellow brown scale at the tip and multiple longitudinal striations [3]. Treatment options include surgical resection, shave excision followed by phenolization, electrofulgration, 1% sirolimus and LASERS [4].

Onychomycosis is a commonly encountered disease of nails. A number of underlying risk factors such as advanced age, obesity, genetic predisposition, diabetes, cancer, immunodeficiency, peripheral arterial disease, tinea pedis, nail damage, and nail psoriasis have been implicated. We believe that nail changes caused by Koenen tumors predisposed to fungal invasion and recurrent infection in our patient. Other cases of nail tumors (onychomatricoma) with concurrent onychomycosis have also been reported [5]. Management can be challenging in these cases and standard antifungal treatment may not suffice. We have planned our patient for shave excision of tumors followed by phenolization along with oral antifungals.

Conclusion

Koenen tumors are well known manifestations of tuberous sclerosis leading to nail changes. Whether this association of onychomycosis with ungual fibromas is a chance occurrence or the splits in nail plate arising due to tumour actually predisposed to recurrent invasion and entry by fungus, needs careful interpretation and larger well controlled studies.

Acknowledgment

None

Conflict of Interest

None

Patient Consent

Written informed consent was obtained from the parents for publication of this case report and accompanying images.

Authorship Statement

Manuscript has been read and approved by all authors, that requirement for authorship as stated have been met and that each author believes manuscript represents honest work.

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Ethics Approval

Not Required.

Data Availability

Research data are not shared.

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