



Exaggerated subungual fibromas in a patient with tuberous sclerosis complex

Tüberoz skleroz kompleksli bir hastada abartılı subungual fibromlar

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To Editor,

Tuberous sclerosis complex (TSC) is an autosomal dominant neurocutaneous condition characterized by hamartomas, which can affect multiple organ systems and cause a wide range of symptoms¹.

A 36-year-old female patient was admitted to the dermatology outpatient clinic with a raised lesion under her fingernails that had been present since she was 18 years old. Dermatological examination revealed skin-colored hard papules under the fingernails (Figure 1A-C) and skin-colored plaques on the lower back (Figure 2). It was discovered that a sibling had similar lesions on the back and under the fingernails. Histopathological examination revealed widespread fibrosis, thin elastic fibers, and collagen bundles extending into the dermis. The biopsy result was consistent with connective tissue nevus. Nail biopsy was recommended, but the patient refused. The primary criteria for the skin manifestations of TSC are nail fibromas and shagreen patches (connective tissue nevus)^{1,2}. Other dermatological findings of TSC, such as sebaceous adenoma, fibrous cephalic plaque, hypopigmented macules, intraoral fibromas, and tooth enamel pits, were not detected.

Although TSC is associated with seizures and mental impairment, these symptoms do not always present. Skin

findings alone can be used to diagnose patients². We diagnosed TSC in this patient because two major criteria were present, and the family history was positive¹. The patient's complete blood count, biochemistry, chest radiography, total abdominal, pelvic, and renal ultrasonography, echocardiography, and brain magnetic resonance imaging findings were normal. Ophthalmological examination revealed no retinal hamartoma. Patients with subungual fibromas commonly complain of this issue, which frequently leads to nail plate distortion, as observed in our patient. Although the therapeutic choices are limited, rapamycin can be administered efficiently and safely in topical applications.³ Topical treatment was not favored in our patient because the lesions were numerous in the subungual region and it was impossible to apply the treatment there, making it impossible to apply the treatment effectively. The patient was advised to undergo surgery; however, she refused. The patient provided written informed consent to publish this case report and accompanying images.

Aldrich et al.⁴ showed that 80% of acral skin lesions in patients with TSC (n=76) had unguinal fibromas. Periungual fibromas were more widespread than subungual fibromas (foot > hand). Periungual fibromas, also known as Koenen tumors, are major criteria for TSC and are almost

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Figure 1. (A, C) Green-brown discoloration, subungual papules, and dystrophic changes in all fingernails; (B) Subungual papules, subungual hyperkeratosis, and pinpoint bleeding in some areas on dermoscopic examination (Dermlite DL4; polarized mode)



Figure 2. Shagreen plaques on both sides of the lower back always present². They begin to appear around puberty and the likelihood of their occurrence increases with age². Subungual fibromas were more frequently found in the fingernails⁴. The middle region of the nail was the most common location for subungual fibromas in both hands and feet⁴. Additionally, most fibromas showed hyperkeratotic changes and pinpoint hemorrhages near the tip. Nails with multiple fibromas also exhibited dystrophic changes⁴.

In our patient, we observed similar hyperkeratotic features and occasional bleeding in subungual fibromas of the hand. Furthermore, all the nails with multiple fibromas exhibited dystrophic nail changes. Long-term pressure damage to the nail matrix is thought to play a role in the formation of longitudinal nail grooves in subungual fibromas, but it has been reported that it can also develop in patients with non-visible periungual fibromas⁴.

In patients with TSC, nail findings may include onychogryphosis, which distorts the nail plate and results in a horn-like appearance, potentially causing pain⁵. Furthermore, noticeable thickening of the nail bed, along with a gradual elevation in the distal nail, which is sometimes accompanied by a roughened nail surface, can be seen⁵. Some nails may exhibit a “pincer” or “omega” appearance, whereas others may become thinner before reaching the fingertip⁵.

Nail fibromas have also been reported as the sole dermatological manifestation of TSC in some patients⁴. The early diagnosis of probable lifetime comorbidities increases the chance of diagnosing potential consequences, thereby improving the patient’s quality of life. Therefore, a multidisciplinary approach is necessary. As TSC can be identified based on skin findings, conducting a complete skin examination, detecting skin symptoms, and raising awareness are crucial actions.

Ethics

Informed Consent: The patient provided written informed consent for the publication of this case report and the accompanying images.

Footnotes

Authorship Contributions

Concept: D.M., E.T.Y., N.K, S.P.K., Design: D.M., E.T.Y., N.K, S.P.K., Data Collection or Processing: D.M., E.T.Y., N.K, S.P.K., Analysis or Interpretation: D.M., E.T.Y., N.K, S.P.K., Writing: D.M., S.P.K.

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