CASE REPORT

Idiopathic 20-nail dystrophy

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SUMMARY

Twenty-nail dystrophy is a rare disease, typically with all 20 nails affected, which normally occurs as an idiopathic condition in childhood but can be linked to other diseases. We report a case of a 7-year-old girl with a 3-year history of 20-nail dystrophy and no associated diseases or family history of skin or nail diseases. She was followed and treated conservatively and, after 6 years of follow-up, we found a marked improvement and almost full resolution of her nail dystrophy. The aim of this report is to show that idiopathic 20-nail dystrophy in children is a self-limiting condition and thus can be treated conservatively.

BACKGROUND

Twenty-nail dystrophy (TND), first described by Hazelrigg *et al*,¹ is an acquired, idiopathic nail dystrophy (trachyonychia) involving all 20 nails.^{1 2} TND is a rare benign condition, normally occurring in childhood, and also reported in adults where lesions tend to persist. The nails are uniformly and simultaneously affected with opalescence, abnormal thickness, fragility and excessive longitudinal ridging.^{1 3 4}

Although TND is often idiopathic and not associated with other diseases, a few case reports have shown possible association between TND and other skin diseases such as eczema, psoriasis, lichen planus, vitiligo and alopecia areata.⁵⁻⁷ A thorough systematic dermatological examination is therefore required to identify possible associated treatable diseases.

The options for treatment of TND are few and not very effective.^{2 8-11} The condition has been shown in the literature to be self-limiting over years, even in the presence of the associated diseases aforementioned, that is why reassurance of the child and parents is essential, to avoid unnecessary, ineffective and potentially harmful medical treatments.²

CASE PRESENTATION

A 7-year-old Caucasian girl was referred to our department of dermatology for a second opinion, with a 3-year history of TND.

At first examination, all 20 nails were thickened and dystrophic with trachyonychia (figure 1A). The girl was otherwise healthy, apart from a congenital hip dysplasia without sequelae and some hypermobility. Especially, she had never had eczema, psoriasis, lichen planus or alopecia areata, and there was no family history of skin diseases or nail dystrophy. Nail scrapings were constantly negative for pathogenic fungi and bacteria, and routine laboratory tests were all normal.

A diagnosis of idiopathic TND was thus maintained.

As the patient did not have any nuisance other than cosmetic discomfort, we chose not to treat her very aggressively. She was treated for some months with calcipotriol topically without any efficacy. A chiropodist took care of the nails in the beginning but after a short period the girl and her family could handle the procedure themselves.

At the age of 13 years, a total of 9 years after initial signs, the girl attended a follow-up visit and showed a marked improvement of the nails with almost total clearance (figure 1B).

DIFFERENTIAL DIAGNOSIS

As a differential diagnosis to TND, pachyonychia congenita and yellow nail syndrome were considered because of the thickness of the nails. We could, however, not confirm these diagnoses, as we found no mucosal involvement or family history nor any signs of lymphoedema or upper airway problems.

Because of the congenital hip dysplasia, hypermobility and a family history of hypermobility (mother and grandmother), hypermobile Ehlers-Danlos syndrome (EDS) was also considered as a differential diagnosis. The patient did not meet diagnostic criteria and nail abnormalities are not a common finding in EDS.

OUTCOME AND FOLLOW-UP

Nine years after initial signs, only conservatively treated, the girl attended a follow-up visit and showed a marked improvement of the nails with almost total clearance (figure 1).

DISCUSSION

Most cases of TND are considered idiopathic. However, a few case reports have shown possible association between TND and other concomitant dermatoses and other comorbidities such as eczema, psoriasis, lichen planus, vitiligo and alopecia areata.^{5 7} In our presented case, there were no associated diseases, although the patient was known to have congenital hip dysplasia and hypermobility.

Some treatment strategies for TND and trachyonychia have been suggested in the literature, such as psoralen plus ultraviolet A (PUVA) phototherapy, glucocorticosteroid injections, tazarotene, cyclosporine and calcipotriol.^{2 8-11}

TND is, in most cases, a self-limiting condition, even in the presence of associated diseases.^{2 3} In our case, we therefore only suggested and tried a mild treatment strategy with topical calcipotriol gel for 3 months, during which time it did not have an

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Figure 1 (A) Twenty-nail dystrophy affecting all fingernails and toenails in a 7-year-old girl. **(B)** Almost total spontaneous remission at the age of 13 years.

effect other than annoying the patient because she could not use nail polish to hide the nail plates.

At times it is difficult to reassure the patient and parents about the nature and natural history of the disease, and they might seek medical treatment to improve the embarrassing nail deformity.

As our case illustrates, we used reassurance and support to form the basis in management of this patient, to avoid unnecessary, ineffective and potentially harming medical therapies. In the end, we saw a marked improvement of the patients' nails with almost total spontaneous remission, which further confirms the diagnosis of self-limiting idiopathic TND.

Learning points

- Twenty-nail dystrophy (TND) is a rare disease, typically with all 20 nails affected, which normally occurs as an idiopathic condition in childhood.
- TND is, in most cases, a self-limiting disease and no medical treatment is needed. However, a careful systematic dermatological examination is important to identify possible associated treatable diseases.
- Reassurance of the child and parents is essential in the management of TND, to avoid unnecessary, ineffective and potentially harmful medical treatments.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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