Dear editor,

The term “idiopathic atrophy of nails” (IAN) was coined by Samman (1969) to describe acquired progressive painless nail loss as the primary cutaneous abnormality in a series of nine patients.[1] Very few cases have been reported so far in the literature.[2-5] Subsequent reports suggested that IAN could be a clinical variant of nail lichen planus, though that remained controversial. A distinction was made from nail atrophy with pterygium as IAN shows nail atrophy with a preserved proximal nail fold (PNF).[6] Even though the original term persists, subsequent reports have associated IAN with a variety of insults. The pathophysiology is quite distinctive and irreversible.

A 12-year-old child presented to us with slowly progressive thinning and loss of nails over the past few years. About 5 years earlier, he had developed an erythematous macular rash with purpuric areas and extensive skin necrosis. It progressed over 24 hours to a sheet-like epidermal detachment with tenderness. Pseudo-Nikolsky’s sign was positive and a diagnosis of toxic epidermal necrolysis (TEN) induced by sulfonamide was made. The child was kept under intensive care and appropriate treatment for about a month. During this period, he developed swelling of multiple nail folds with subsequent onychomadesis. There was extensive involvement of the ocular mucosa with erosions and ulceration. Subsequent healing of skin, eyes, and nails was a prolonged and slow process, with progressively thinner nails, leading to the current status.

On examination, he had marked thinning, atrophy, frayed edges, and koilonychia involving all the nails [Figure 1a, 1b]. However, all the nail folds were normal without any signs of pterygium or scarring. Few hyperpigmented and atrophic scars involving face and trunk were also noticed. The child had extensive synchiae and corneal opacities with near-complete loss of vision in both eyes (only perception of light). Based on the history and examination, a diagnosis of IAN was made. The parents did not consent for nail biopsy.

Clinically, IAN presents as slow, painless, progressive atrophy of some or all the nails. Initial cases reported by Samman had pterygium formation in a few nails[1] [Table 1]. Subsequent cases by Colver and Dawber demonstrated lichen planus as a common histopathological finding,[3] leading to the conclusion that childhood IAN could be a variant of lichen planus. Two years later, Barth et al.[2] reported four cases where IAN was not associated with pterygium, and none of the cases demonstrated lichen planus on histopathology. They proposed that IAN could be the end result of matrix damage due to trauma, chemical injury, lichen planus, ischemia, etc., This appears to be the most plausible explanation in our case. Barth et al.[3], concluded that atrophy of nails with preservation of nail folds signifies IAN, while atrophy with pterygium signifies lichen planus. The name “idiopathic atrophy” had been retained over the years, even though subsequent reports have attributed varied causes, as was seen in our case as well. It could be reasonably concluded that IAN is in fact a common end point for a number of significant matrix insults. As IAN develops slowly and progressively, the cause may go unrecognized in some cases.

Suarez and Scher proposed a possible genetic basis for IAN due to co-occurrence in siblings.[4] Cases reported by Tosti et al.[5] had nail atrophy with pterygium in a few of their nails.[6] This histopathological study proposed that IAN is characterized by nail matrix hypergranulosis with disappearance of keratogenous zone, which may be the final result of varied inflammatory or compressive insults to the nail matrix including lichen planus, acrodermatitis continua of Hallopeau, systemic amyloidosis, etc., Tosti et al.[5] also proposed that cell differentiation progressing vertically instead of diagonally leads to displacement and uplifting of the PNF in this condition. This results in an increase in the nail matrix angle. As a result, the distal nail plate appears considerably thinner and irregular. A compact horny layer replaces the abnormal dorsal nail plate which desquamates shortly after its emergence from under the PNF. As the apparent nail plate is composed of this horny layer only, it appears frayed, disintegrated, and excessively curved (koilonychia). Thus, IAN is a pathological end point irrespective of the inciting etiology including nail lichen planus, acrodermatitis continua, systemic amyloidosis, pemphigus, atopic eczema, etc., The normal nail matrix keratinization may never be restored even when the insult ceases, and keratinization by granular cells may persist indefinitely causing a permanent nail plate atrophy.

In our case, the clinical findings and lack of other cutaneous and mucosal lesions suggested IAN; however, history clearly outlined TEN as the initiating insult. Our report highlights that IAN may not always be “idiopathic.” It should be considered as a morphological end point of severe nail matrix insult, causing irreversible damage to nail matrix keratinization with a preserved nail-fold architecture; hence, it can be better labelled as ‘progressive atrophy of nails’
Table 1: Characteristics of cases with “idiopathic atrophy of nails” reported in literature

| Year and authors          | Number of patients | Clinical characteristics                                                                 | Histological features                                                                 |
|---------------------------|--------------------|------------------------------------------------------------------------------------------|----------------------------------------------------------------------------------------|
| Samman (1969)[1]          | 9                  | Features ranging from discoloration, ridging, pterygium, and permanent total painless nail loss with obliteration of the nail fold | Not evaluated                                                                          |
| Colver and Dawber (1987)[3] | 4                  | Longitudinal ridging, nail atrophy, and pterygium with nail-fold scarring in one case | Longitudinal nail biopsy- suggestive of active lichen planus in 3 cases and scarring lichen planus in 1 case |
| Barth et al. (1988)[2]    | 4                  | Longitudinal ridging, atrophy of nail plate, and mild pterygium in one case Koilonychia in one case Preservation of nail folds Atopic eczema in one case | Longitudinal nail biopsy—none of the cases showed features of lichen planus Abnormal maturation of nail bed epithelium was a consistent feature |
| Suarez and Scher (1990)[4] | 3                  | Two siblings with 20 nail involvement with longitudinal ridging, discoloration, splitting of nail plate, atrophy, and nail bed scarring Third case had longitudinal ridging and early atrophy of nail plate with frayed distal edges | Not evaluated                                                                          |
| DeBerker et al. (1993)[6] | 2                  | Trachyonychia, discoloration, nail splitting, longitudinal ridging, onycholysis, proximal nail thickening, and atrophy | Nail matrix biopsy suggestive of pemphigus vulgaris with suprabasal split and acantholysis; DIF positive for IgG in intercellular pattern; IIF positive for IgG |
| Tosti et al. (1995)[5]    | 2                  | Severe nail plate thinning without obliteration of proximal nail fold Atrophy of nail plate with scarring and pterygium One case had prolonged history of Raynaud’s phenomenon | Longitudinal nail biopsy: not diagnostic, showing nail matrix hypergranulosis with absence of keratogenous zone |
| Tosti et al. (2001)[7]    | 3                  | Severe nail atrophy of all 20 nails with and without pterygium formation (scarring with pterygium), severe nail thinning without obliteration of PNF | Longitudinal nail biopsy: nail matrix hypergranulosis, absence of keratogenous zone, mild band-like lymphocytic infiltrate in dermis of PNF (one sample) |

DIF=Direct immunofluorescence, IIF=indirect immunofluorescence, PNF=proximal nail fold

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Conflicts of interest
There are no conflicts of interest.

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