Pterygium of the Nail

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GOAL

To summarize the clinical features and etiologies of pterygium of the nail

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Describe the clinical features of pterygium of the nail.
2. Discuss the pathogenesis of both dorsal pterygium and ventral pterygium.
3. Identify the causes of dorsal and ventral pterygia.

CME Test on page 347.

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Dorsal pterygium is the growth of a projection of the proximal nail fold that fuses with the underlying matrix and, subsequently, with the nail bed. As a result, the nail plate is divided into 2 lateral segments. Ventral pterygium, a distal extension of the hyponychium that anchors to the undersurface of the nail plate, eventually obliterates the distal nail groove.

Dorsal Pterygium

Dorsal pterygium, or pterygium unguis, occurs when the cul-de-sac of the proximal nail fold (PNF) grad-

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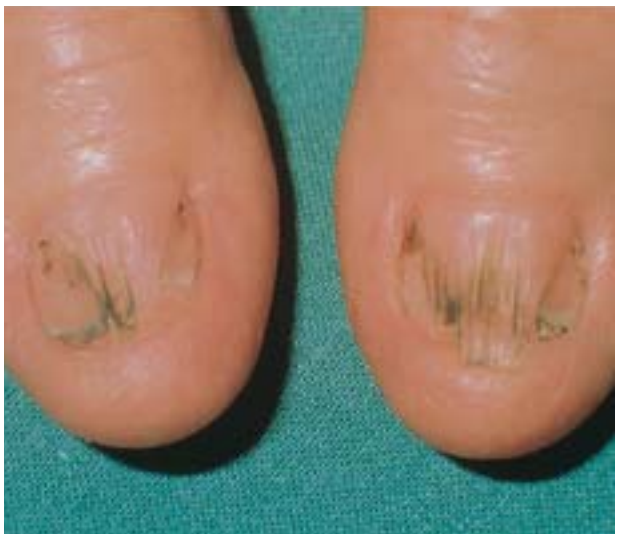

FIGURE 1. Lichen planus involving both thumbs; lateral remnants of nail keratin are visible.

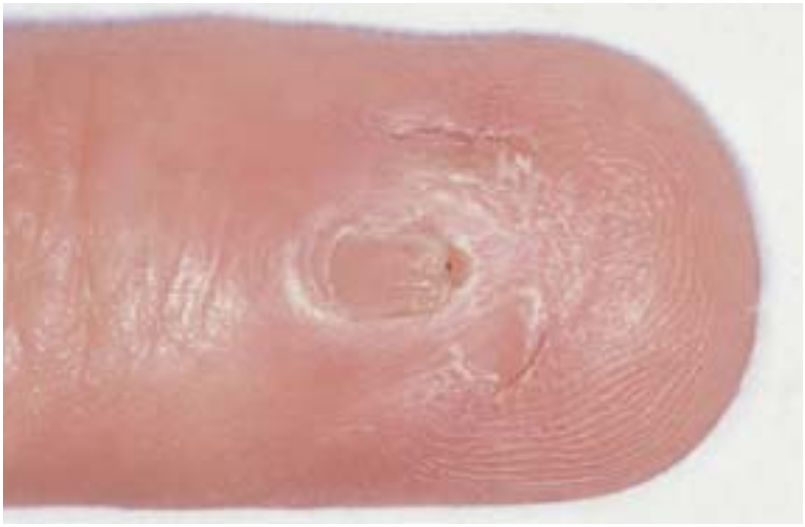

FIGURE 2. Dorsal pterygium involving one finger after a major trauma.

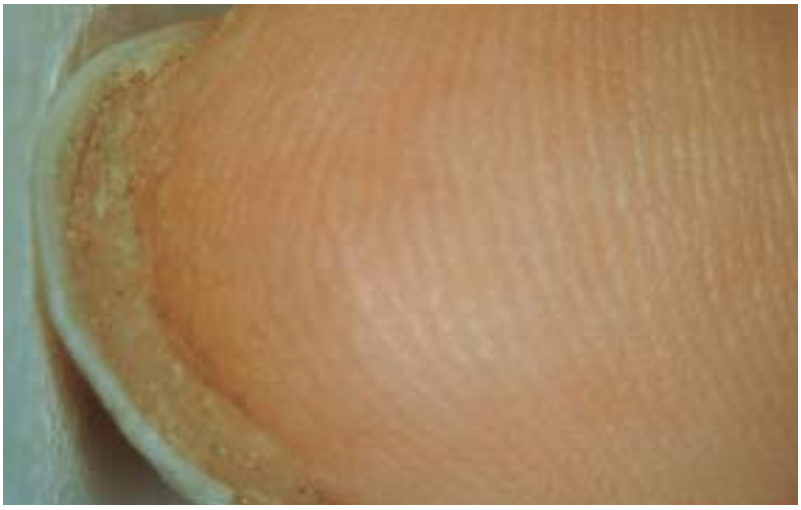

FIGURE 3. Ventral pterygium affecting only one finger.

ually shortens and the nail plate thins until, eventually, the PNF fuses to the matrix and, subsequently, to the nail bed. The resulting nail plate segments progressively decrease in size as the pterygium widens, resulting in 2 small remnants involving the PNF. Complete involvement of the matrix and nail bed produces a total loss of the plate and permanent atrophy of the nail apparatus. Dorsal pterygium is usually acquired, but exceptional congenital forms have been reported. In a case of nail lichen planus in 1921, Friedman first described pterygium unguis using the term *navellierungsprozess*.[1] Dorsal pterygium rarely affects the toes; involvement of all 20 toenails has been reported in only one case.

Although pterygium unguis is most commonly caused by lichen planus (Figure 1),[2] it also may originate from burns, radiodermatitis, trauma (Figure 2), and diseases prone to developing adherence bands, including cicatricial pemphigoid, graft versus host disease, toxic epidermal necrolysis, and pemphigus foliaceus (Table I). Peripheral ischemia, intermittently detected in Raynaud's phenomenon[3] or permanently found in atherosclerosis, diabetic vasculopathy, and type 2 lepra reaction, has been identified as a vascular cause of dorsal pterygium. An idiopathic form of pterygium unguis, or idiopathic atrophy of the nails, exists, but may be a variation of lichen planus and remains a controversial cause.[4] Congenital forms may be associated with dyskeratosis congenita, and pterygium formation has been reported in systemic lupus erythematosus and in one case of sarcoidosis involving the PNF.

Healing of a disease involving the PNF may lead to scarring pterygium formation; however, several factors that interfere with the healing of the PNF must appear simultaneously for this to occur. In patients with dorsal pterygium (excluding the traumatic and congenital types), the main factor is dilatation of the nail capillary loops and the formation of a slender, microvascular shunt system in them. Although pterygium that

Table I.

Causes of Dorsal Pterygium (Pterygium Unguis)

Atherosclerosis
Burns
Cicatricial pemphigoid
Congenital etiology
Diabetic vasculopathy
Dyskeratosis congenita
Graft versus host disease
Idiopathic atrophy of the nails[3]
Inadequate corticosteroid matrix infiltration for *Candida paronychia*
Lichen planus[1]
Onychotillomania
Pemphigus foliaceus
Radiodermatitis
Raynaud's phenomenon[3]
Sarcoidosis involving the PNF
Systemic lupus erythematosus
Toxic epidermal necrolysis
Trauma
Type 2 lepra reaction

Table II.

Causes of Ventral Pterygium (Pterygium Inversum Unguis)

Causalgia of the median nerve
Congenital etiology
Family history
Formaldehyde-containing hardeners[8]
Lenticular atrophy of the palmar creases
Leprosy
Neurofibromatosis
Paresis
Scarring in the vicinity of the distal nail groove
Subungual exostosis
Systemic connective tissue diseases
Systemic lupus erythematosus
Systemic sclerosis

results from trauma is not linked to the intensity of the trauma, it may be observed in severe distal injury. Also, pterygium remains exceptional in repeated chronic trauma inflicted to the PNF in onychotillomania, and we observed one pterygium unguis arising after inadequate corticosteroid matrix infiltration for *Candida paronychia* (Richert, unpublished data, 1997).

Ventral Pterygium

Ventral pterygium (Figure 3) is a newly described condition in which a distal extension of the hyponychium anchors to the undersurface of the nail plate and, subsequently, obliterates the distal nail groove. The condition was first described in 1973 by Caputo and Prandi,[5] who coined the term *pterygium inversum unguis* (PIU) to describe a forward extension of the hyponychium anchoring to the undersurface of the nail plate and, thus, obliterating the distal nail groove.

PIU may be either congenital or acquired. Odom et al[6] first described the congenital form as a "congenital, painful and aberrant hyponychium" in 1974. In some instances, PIU has been reported as familial and, in most reported cases, patients sought medical advice for the pain or bleeding they experienced when trimming their nails. Causes of PIU are listed in Table II.

Because the frequency of ventral pterygium may be underestimated, especially in asymptomatic cases, Mello Filho[7] performed a systematic digital examination on 2000 patients and found that 0.4% of the adult population was affected. In these cases there was no family history, and the condition was observed more frequently in women than in men. Idiopathic forms were preponderant, and involvement of the toes remains exceptional.

Acquired PIU, which occurs in 16% of patients, is by far the most common form and, though idiopathic, is usually secondary to systemic connective tissue diseases, progressive systemic sclerosis, and systemic lupus erythematosus. One patient with congenital PIU developed systemic lupus erythematosus at the age of 19, but whether this was coincidental

FIGURE 4. Ventral pterygium of several digits after applications of nail polish containing formaldehyde.

or related has not been determined.

PIU may be secondary to scarring in the vicinity of the distal nail groove, or it may arise from a reaction to formaldehyde-containing nail hardeners[8] (Figure 4) or from subungual exostosis. PIU has been reported once in neurofibromatosis, which was associated with lenticular atrophy of the palmar creases and causalgia of the median nerve. Unilateral PIU of the fingers and toes has been reported 1 year after a stroke and resulted in paresis of the same side.

Comments

Pterygium of the nail has been described on both dorsal and ventral aspects of the nail plate. The dorsal pterygium consists of a forward growth of a projection of the proximal nail fold that fuses with the underlying matrix and, subsequently, with the nail bed and divides the nail plate in 2 lateral segments. Ventral pterygium is a distal extension of the hyponychium that anchors to the undersurface of the nail plate and eventually obliterates the distal nail groove. Both conditions are nonspecific abnormalities of the nail apparatus.

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