Inflammatory Diseases of the Nail: Clinical Overview

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Contents

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16.1 Introduction

Diseases of the nail and its adjacent tissues rank among the most interesting and fascinating diseases that "flesh is heir to." Diagnosis for these diseases may require examinations such as histology, hematology, radiography, magnetic resonance imaging, and scintigraphy. Advancing technology has recently taken a leap forward with the development of 2D and 3D ultrasound. This development in investigative techniques combines ease of use with economy of cost.

16.2 Psoriasis

Five per cent of patients with nail psoriasis do not show any skin involvement [1].

The histology of psoriasis is an inflammatory parakeratotic papule. This same inflammatory papule occurs in the nails of psoriatic patients and the location of the papule determines the clinical presentation [2].

Erythematous scaly papules and plaques can be seen on the cutaneous surface of the proximal nail fold (PNF) and hyponychium. Their appearance is similar to psoriasis in other areas of the skin. A psoriatic lesion of the undersurface of the PNF produces a separation of the PNF from the nail plate with a secondary paronychia.

Psoriatic lesions can occur in the proximal portion of the matrix or in the distal matrix. A cluster of abnormal parakeratotic cells is incorporated into the dorsal surface of the nail if the lesion is present in the proximal matrix. These loosely arranged cells might travel with the nail plate for a short distance but are lost quickly, and pits (Fig. 16.1) mark the prior location of these cells. If the parakeratotic cells are in the central or distal portion of the matrix, they are "trapped" within the nail plate and reflected light gives the nail plate an opaque white appearance.

Psoriatic lesions can occur in the nail bed. The subungual area can frequently present with hyperkeratosis (Fig. 16.2) or

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Fig. 16.1 Nail pitting

Fig. 16.2 Onycholysis with proximal erythematous border

onycholysis, whereas typical cases are proximally surrounded by an erythematous line (Fig. 16.3). A deeply erythematous macule can be seen through the translucent nail plate. Psoriatic lesions produce glycoproteins that accumulate under the nail and present clinically with the "oil dropping"' sign.

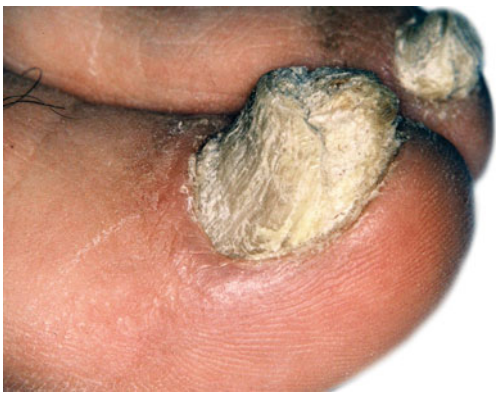

Fig. 16.3 Subungual hyperkeratosis

Fig. 16.4 Transverse leuconychia of the thumb

A constellation of the findings described here is usually seen, depending on the location of the psoriatic lesions. A completely dystrophic nail characterized by mounds of keratinaceous debris can be seen if the psoriatic lesion is particularly large, especially in pustular psoriasis or acrodermatitis continua of Hallopeau. The severity of nail disease can be correlated with skin disease that is severe and psoriatic arthritis that is advanced [3–8].

Intermittent inflammation of the nail matrix can produce Beau's lines. Other less common nail changes described in psoriasis are nail fold telangiectasias, red lunulae, punctuate red spots in the lunula, transverse leukonychia (Fig. 16.4), and leukonychia punctata.

Ten to twenty percent of patients with psoriasis present with psoriatic arthritis (PsA), and patients with PsA commonly have nail involvement (53–86 %).

Psoriatic onychopathy may present varying degrees of alterations on ultrasound in both the nail bed and the nail

plate. These abnormalities (from early to late phases) are the thickening of the nail bed (increased distance between the ventral plate and the bony margin of the distal phalanx); increased blood flow within the nail bed; loss of definition of the ventral plate; hyperechoic focal involvement of the ventral plate (that can be subclinical) usually correlating with the subungual keratosis and can present without involvement of the dorsal plate. Thickening, loss of definition, and undulation of both nail plates can be detected in the late phases.

The assessment of activity in psoriatic onychopathy is possible using sonography. On color Doppler ultrasound, registration of the basal activity (blood flow and sonographic anatomical changes) can be compared with the modifications detected on a follow-up examination.

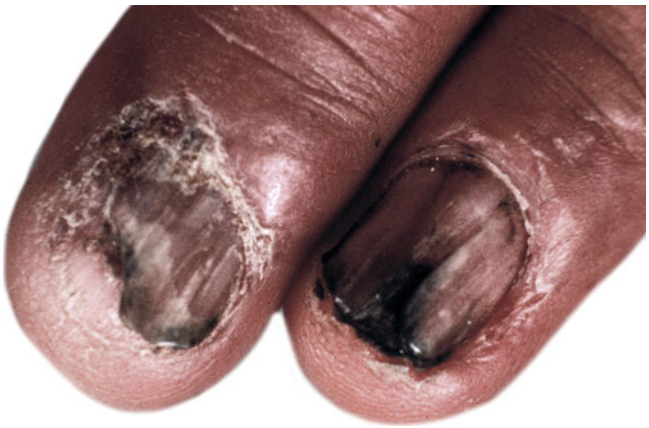

Fig. 16.5 Discoid type of lupus erythematosus (Courtesy B. Richert, Belgium)

16.3 Lupus Erythematous

Lupus erythematous is a multisystemic connective tissue disease, characterized by the presence of numerous auto-antibodies, circulating immune complexes, and widespread immunologically determined tissue damage [9, 10] (Figs. 16.5 and 16.6).

16.3.1 Arthritis, Arthralgia, and Fever

Arthritis can be transient, migratory, or a more persistant seronegative polyarthritis.

16.3.2 Skin Lesions

Skin lesions are seen in more than two thirds of patients. In addition to the classic, photosensitive, erythematous butterfly rash across the face, a vasculitic rash can present as purpura or periungual erythema with "chilblain-like" lesions or digital infarcts.

Cardiopulmonary features, renal, and central nervous system involvement can be associated with the joint and cutaneous signs.

Interestingly, blood flow abnormalities that involve the digital arteries and nail bed can be detected using color Doppler ultrasound. These changes include thrombotic phenomena within the distal end of the digital arteries that can cause secondary hypovascular changes in the nail bed leading to dystrophic changes. Thus, variable areas of thickness can be seen in the nail bed, together with thickening and thinning areas associated with this secondary dystrophy that can be accompanied by a discontinuity of the nail plates.

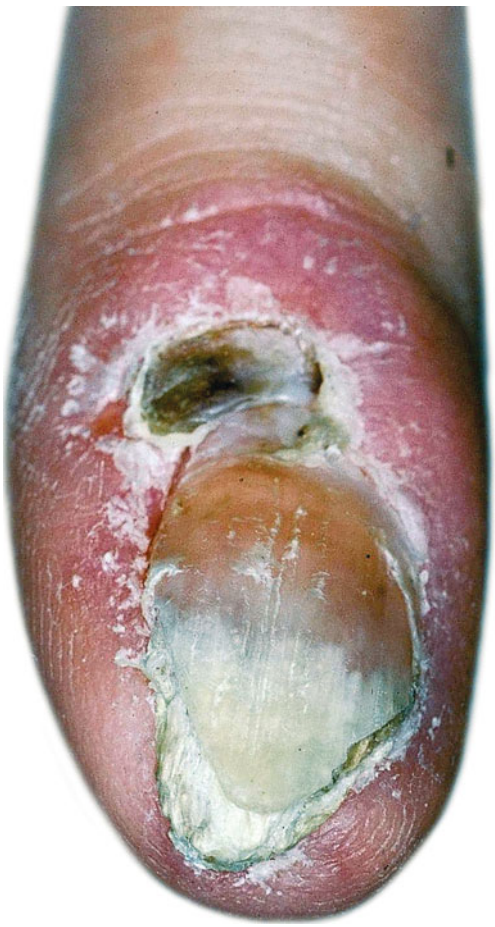

Fig. 16.6 Ulceration of the proximal nail fold in lupus erythematosus (Courtesy B. Richert, Belgium)

16.4 Scleroderma

Initially, there is often well-demarcated non-pitting oedema and induration associated with 'sausage' swelling and restriction of movement of the fingers seen in scleroderma [11]. Later, the skin becomes shiny, with atrophy and ulceration of the fingertips, with or without associated calcinosis. The skin of the face, limbs, and torso is variably affected and there can be striking pigmentation and telangiectasia. As the disease progresses, the face can become taut and mask-like, with 'beaking' of the nose and difficulty in opening the mouth. The tightening of the skin over bony prominences results in flexion contractures and liability to trauma. Often there is erythema and dilated capillaries of the PNFs.

Systemic sclerosis can affect both microvascular structures and functions of the nail unit. These changes have been previously registered by using other imaging modalities such as laser, Doppler, thermal imaging, and nailfold capillaroscopy. Giant capillaries, hemorrhages, and/or avascular areas have been described in scleroderma using such imaging modalities. Thus, vascular abnormalities are one of the primary pathologic components of scleroderma (Figs. 16.7 and 16.8).

Thickening and decreased echogenicity of the nail bed with an upward displacement of the nail plates can be seen using ultrasound, and is probably related to edema and/or chronic inflammatory changes. Hypovascularity of the nail bed is sometimes detected and can be related to changes at the microvascularity level.

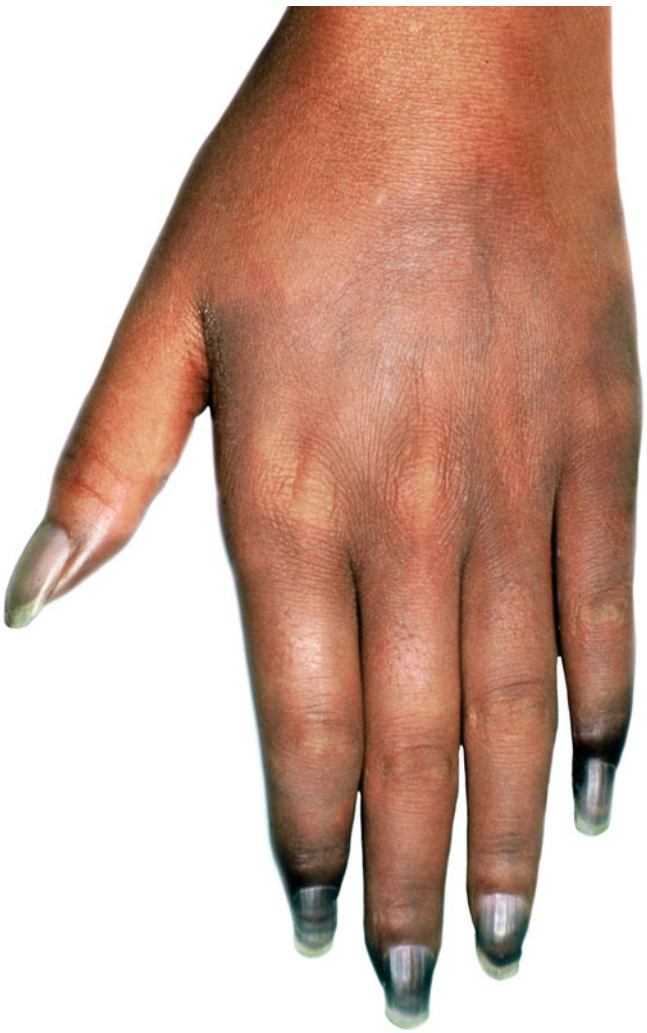

Fig. 16.7 Systemic sclerodermia with vascular impairment of the distal digits

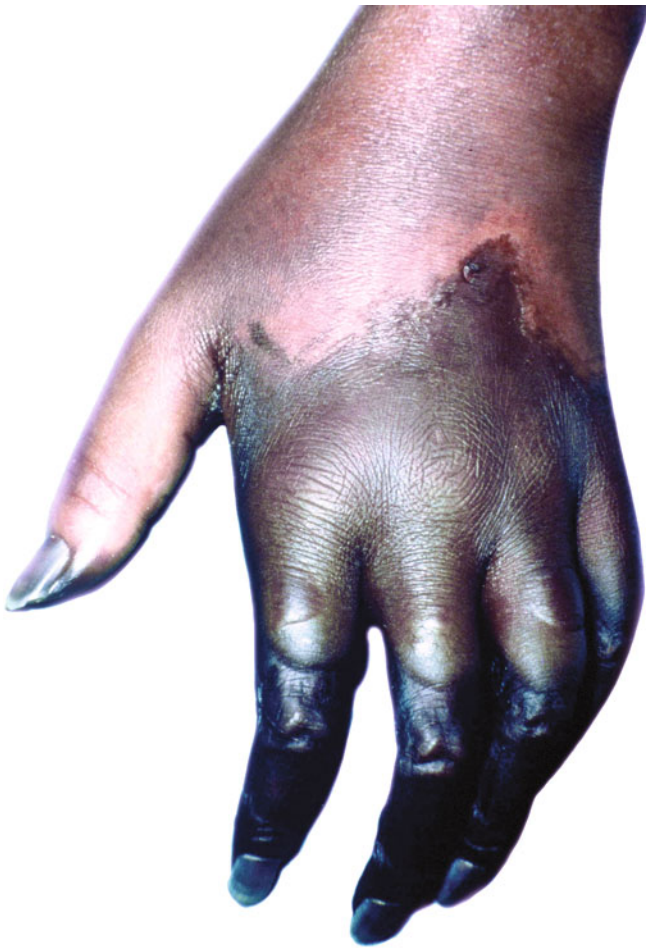

Fig. 16.8 Same patient as in Fig. 16.7, late stage (Courtesy N. Rowell)

16.5 Dermatomyositis

16.5.1 Adult Dermatomyositis [11]

Adult dermatomyositis is more common in women than in men. Acute or subacute muscle weakness is accompanied by periorbital edema and a characteristic violet "heliotrope" rash on the upper eyelids. Additionally, a photosensitive, erythematous, scaling rash on the face, shoulders, upper arms, and chest with red patches over the knuckles, elbows, and knees can be seen. Muscle pain, tenderness, and weight loss are common, as are arthralgia and mild inflammatory polyarthritis.

Inflammatory myositis can be associated with malignancy.

16.5.2 Childhood Dermatomyositis [11]

Childhood dermatomyositis most commonly affects children between the ages of 4 and 10 years old. Muscle weakness is usually accompanied by the typical rash of dermatomyositis and muscle atrophy, contractures, and subcutaneous calcification can be widespread and severe.

Many periungual capillary changes have been described in active juvenile dermatomyositis, among them dilated and tortuous blood vessels, areas of atrophy, telangiectases, central areas of hemorrhages, splinter hemorrhages, and bushy capillary loop formation in the PNF.

Color Doppler ultrasound may register vascularity changes in the nail bed and also detect periungual calcinosis deposits that are frequently located in the fingertips. Calcinosis appears on ultrasound as hyperechoic focal deposits frequently presenting posterior acoustic shadowing typical of the calcium component.

16.6 Rheumatoid Arthritis

Rheumatoid arthritis (RA) is the most common form of chronic inflammatory joint disease. In its typical form, RA is a symmetrical, destructive, and deforming polyarthritis affecting small and large synovial joints, with associated systemic disturbance, a variety of extra-articular features, and the presence of circulating antiglobulin antibodies (rheumatoid factors).

RA is a chronic progressive disorder characterized by symmetric inflammatory arthritis in association with systemic symptoms. Although considered a "joint disease," RA is associated with involvement in diverse organ systems, including the skin and nails. Clinical nail abnormalities that are commonly described that are associated with RA are longitudinal ridging and clubbing [11].

Identification of anatomic changes in the joint, bony margins, tendons, and soft tissues can be visualized using color Doppler ultrasound. The alterations include narrowing of the joint space, tendinosis (degeneration of the fibrillar pattern of the tendon), tear or atrophy of extensor and flexor tendons, erosions of the bony margin, periarticular and peritendinous edema, thickening decreased echogenicity, and increased blood flow in the nail bed.

16.7 Lichen Planus

The etiology of lichen planus is unknown, there is however, some evidence for a genetic susceptibility. Primary immunological disturbance is another likely hypothesis. Nail involvement of one or all the nail components occurs in 10 % of patients with lichen planus, and clinical features depend on the site affected by the pathological process [2].

16.7.1 Nail Fold Involvement

The dorsum of the PNF can be blue or red with or without swelling, and indicates that the proximal matrix is the origin of the lesion and that nail plate changes are likely to occur soon afterward.

16.7.2 Matrix Disease

A small focus of lichen planus in the matrix can present as a bulge under the PNF. The nail gradually reflects the disease process with a longitudinal red line indicating a thinned nail plate evolving into a distal split. The lunula can also be red, either in a focal or generalized pattern, and the next stage is a complete split. The matrix disease is relatively advanced and there can be pterygium formation between the underlying matrix disease and the overlying PNF. The most drastic form of matrix disease is seen in ulcerative lichen planus (Fig. 16.9) where complete, and sometimes irreversible, nail

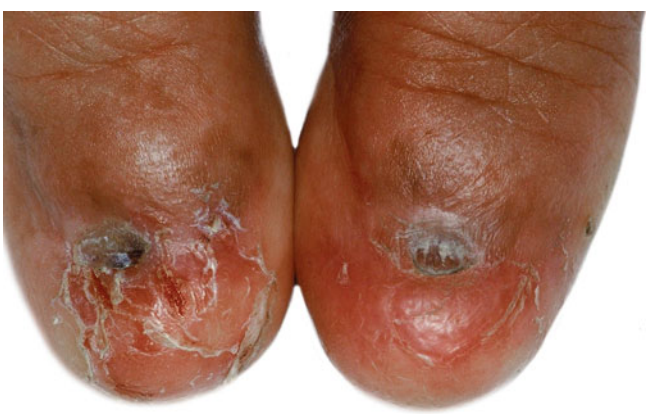

Fig. 16.9 Ulcerative lichen planus

loss combines with large areas of bullae formation and erosion, usually on the soles of the feet and sometimes on the palms of the hands.

Focal disease that does not proceed to significant scarring may leave pigmentary changes similar to lesions seen on the skin. Longitudinal ridging can be a manifestation of lichen planus where disease matrix involvement results in selective atrophy of the nail plate.

16.7.3 Nail Bed Disease

Lichen planus seldom exclusively involves the nail as subungual hyperkeratosis or onycholysis.

16.7.4 Prognosis

The prognosis of the disease depends on the degree of matrix involvement and scarring with pterygium formation.

16.8 Lichen Striatus

Lichen striatus is a linear dermatosis of unknown etiology. It is characterized by the sudden appearance of erythematous, squamous, or lichenoid papules arranged on a continuous or interrupted streak involving the entire length of an extremity. It can extend along a finger or a toe as far as the PNF and affect the nail plate [12]. Several types of nail dystrophy include fraying, longitudinal splitting, punctuate or transverse leuconychia, shredding, onycholysis, and total nail loss. The nail dystrophy may precede the onset of the rash and an isolated assymetrical nail dystrophy in a young person should raise suspicion about the diagnosis.

16.8.1 Prognosis

All these lesions are transient, usually resolving in 1 year. However, the presence of nail involvement indicates a protracted course and the deformity of the nail plate may persist for several years.

16.9 Retronychia

Retronychia is a new pattern of ingrown nail that is characterized by a proximal embedding of the nail plate. De Berker et al. [13, 14] presented a series of 10 cases with a mild-to-moderate paronychia (Fig. 16.10) as a result of 2–4 nail plates being superimposed on each other. Pain, inflammation, and varying degrees of granulation tissue reaction are observed. These symptoms subside rapidly after avulsion. A transitory growth arrest produces a Beau's line, and if the arrest ranges from 3 to 8 weeks, the nail plate separates from the subungual tissues leading to latent onychomadesis with secondary nail shedding [15].

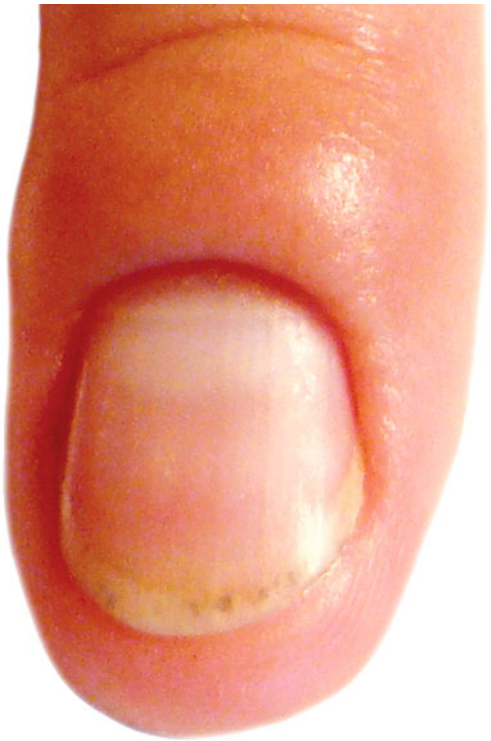

Fig. 16.10 Finger retronychia (Courtesy X. Wortsman)

Important sonographic criteria for diagnosing retronychia are the reduced distance between the origin of the nail plate and the base of the distal phalanx (level of the distal interphalangeal joint compared with the contralateral digit), thickening; decreased echogenicity and increased blood flow in the dermis of the posterior nail fold and proximal nail bed (color Doppler ultrasound).

References

1. Baran R. The burden of nail psoriasis: an introduction. Dermatology. 2010;221 Suppl 1:1–5.
2. Holzberg M. Common nail disorders. Dermatol Clin. 2006;24:349–54.
3. Scarpa, Manguso F, Oriente A, Peluso R, Atteno M, Oriente P. Is the involvement of the DIP joint in psoriatic patient related to nail psoriasis? Clin Rheumatol. 2004;23:27–30.
4. McGonagle D, Ash Z, Dickie L, McDermott M, Aydin SZ. Ann Rheum Dis. 2011;Suppl 1:i71–6.

5. McGonagle D. Enthesis an autoinflammatory lesion linking nail and joint involvement in psoriatic disease. J Eur Acad Dermatol Venereol. 2009;1:9–13.
6. McGonagle D, Benjamin M, Tan AL. The pathogenesis of psoriatic arthritis and associated nail disease: not autoimmune after all? Curr Opin Rheumatol. 2009;21:340–7.
7. McGonagle D, Tan AL, Benjamin M. The nail as a musculoskeletal appendage-implications for an improved understanding of the link between psoriasis and arthritis. Dermatology. 2009;218:97–102.
8. McGonagle D, Tan AL, Benjamin M. The biomechanical link between skin and joint disease in psoriasis and psoriatic arthritis: what every dermatologist needs to know. Ann Rheum Dis. 2008;67:1–4.
9. Kanvar J, Sandipan D, Srabani G. Involvement of nails in discroid. LE JAP I. 1993;41:543.
10. Richert B, André J, Bourguignon R, de la Brassine M. Hyperkeratotic nail discoid lupus erythematosus evolving forwards systemic lupus erythematosus: therapeutic difficulties. J Eur Acad Dermatol Venereol. 2004;18:728–30.
11. Braverman I. Connective tissue (rheumatic) diseases, chapter 7. In: Skin signs of systemic disease. 3rd ed. Philadelphia/London: W. Sanders; 1998. p. 198–277.
12. Baran R, Dupré A, Lauret P, Puissant A. Lichen striatus with nail involvement. Report of 4 cases and review of the 4 cases in the literature. Ann Dermatol Venereol. 1979;106:885–91.
13. De Berker DA, Rendall JR. Retronychia-proximal ingrowing nail. J Eur Acad Dermatol Venereol. 1999;12 Suppl 2:S126.
14. De Berker DA, Richert B, Duhard E, et al. Retronychia: proximal ingrowing of the nail plate. J Am Acad Dermatol. 2008;58: 978–83.
15. Wortsman X, Calderon P, Baran R. Finger retronychias detected early by 3D ultrasound examination. J Eur Acad Dermatol Venereol. 2012;26(2):254–6.