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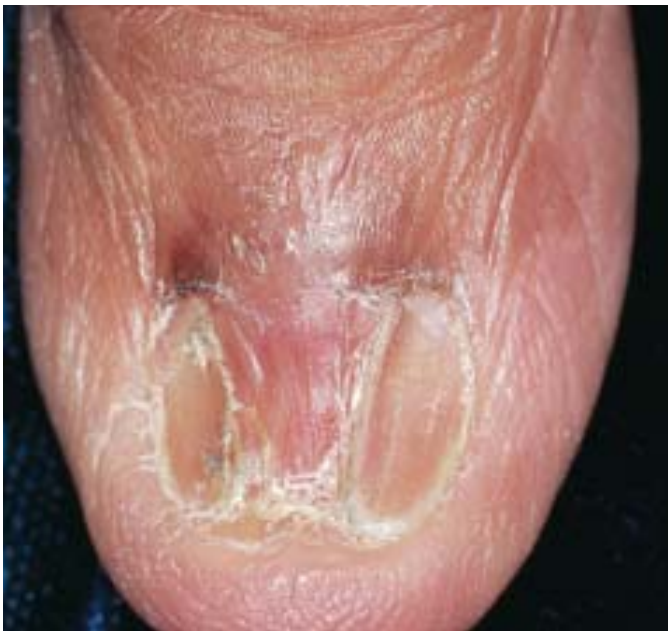

A patient presented with a rapidly progressive deformity of the nails.

What is your diagnosis?

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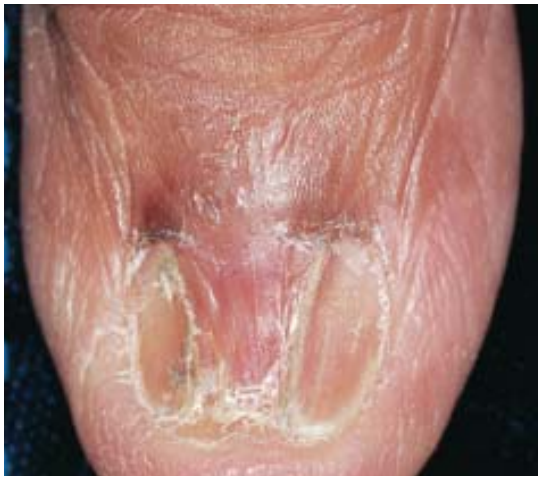
The Diagnosis: Lichen Planus of the Nail



Lichen planus of the nail can be rapidly destructive, resulting in pterygium formation, as in the present case. Significant functional disability is associated with permanent nail damage. Tasks as simple as picking up a penny become nearly impossible. The cosmetic deformity that accompanies the condition is often quite distressing for patients. Permanent anonychia also may occur.[1]

The manifestations of lichen planus of the nail unit present a wide spectrum of clinical disease, depending in part on the portions of the nail unit involved.[2] Most patients are not severely affected, and therapy should be tailored to the extent of disease. Tosti et al[3] reported an excellent response to therapy in a series of 24 patients. Only 25% of their patients had associated lichen planus at other sites, and all patients had a low incidence of severely destructive nail disease.[3] When lichen planus occurs, severely destructive nail disease is often rapidly progressive and can be resistant to therapy. Oral corticosteroids at doses of at least 1 mg/kg per day are generally effective in inducing a temporary remission of rapidly destructive disease. Intramuscular injections of triamcinolone can produce long-term remission in some patients. Complications of long-term systemic corticosteroid therapy can limit its duration.

Alternative therapeutic options are not uniformly successful. Intralesional injection of a corticosteroid such as triamcinolone is often effective and reduces the risk of systemic corticosteroid complications. Digital local anesthetic blocks can be quite helpful in allowing adequate infiltration of the nail matrix and proximal nail bed. Patients are usually far more accepting of intralesional therapy when it is done in conjunction with a digital block. Potent topical steroids represent an alternative to injections, but the response is unpredictable, and topical agents are generally inadequate to treat rapidly progressive severe disease. Alternative therapeutic agents, such as retinoids, antimalarial agents, immune modulators, and immunospressive agents, such as cyclosporine and mycophenolate mofetil, require further study. Griseofulvin has been used in India for the treatment of lichen planus, including severe nail disease.[4] However, I have not experienced therapeutic success with griseofulvin in lichen planus of the nail.

Biopsy findings in affected nails vary during the course of the disease. Active disease is characterized by a lichenoid inflammatory infiltrate with destruction of basal keratinocytes. The nail matrix often exhibits a granular layer, a finding not noted in the normal matrix. Keratohyaline granules in nail matrix biopsies are a nonspecific finding associated with a variety of inflammatory and traumatic insults to the matrix.[5] Because the cause of lichen planus is currently unknown, it is possible that it represents a heterogeneous group of disorders that share a histologic reaction pattern.

Twenty-nail dystrophy, presenting as rough nails (trachonychia), also may be a manifestation of lichen planus.[6,7] Some patients manifest

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other systemic or skin and skin-appendage abnormalities such as alopecia universalis, hypogammaglobulinemia, and ichthyosis vulgaris.[8] Lichen planus of the nail is sometimes associated with unusual cutaneous features such as bullous and ulcerative lichen planus.[9] An association of unusual lichen planus variants with viral hepatitis, especially viral hepatitis type C, has been noted in some cases. Idiopathic atrophy of the nails in children also may be a manifestation of lichen planus.[10] Rarely, hyperkeratotic lesions of lichen planus can mimic tumors of the nail bed.[11] In cases of acquired nail dystrophy where biopsy confirmation of the diagnosis is required, longitudinal nail biopsy provides a good diagnostic yield.[12]

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