Localized Longitudinal Erythronychia

Diagnostic Significance and Physical Explanation

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Background: Longitudinal erythronychia (LE) is a term for red streaks in the nail. We describe the range of diseases manifested by localized (single or bifid) LE and explain the underlying physical changes.

Observations: Longitudinal erythronychia can be multiple or localized. Multiple lesions typically indicate an inflammatory disease such as lichen planus. When localized, they may be a single or bifid streak arising through a benign or malignant neoplasm, scarring of the dermis or epidermis, or the first stage of an inflammatory process that may evolve into multiple LE. Excision of a localized LE may provide a diagnosis and cure. Incisional matrix biopsy of multiple LE may provide a diagnosis. Clinical manifestation of LE arises through reduced compression of the nail bed due to loss of bulk of the nail plate with a groove on the undersurface. A streak of thinned nail then allows an enhanced view of a corresponding streak of engorged nail bed. The reduction in nail thickness renders it more fragile with a tendency to split distally.

Conclusions: Understanding LE can assist in diagnosis and explanation to the patient. Localized LE may represent a focal tumor or dysplastic process.

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For the purposes of this article, we define longitudinal erythronychia (LE) as a longitudinal streak or band in the nail plate commencing from within the matrix and running to the point of separation of the nail bed and nail plate.1 This point is known as the onychocorneal band and represents a zone of specialized attachment between nail and nail bed. This zone is important in the prevention of onycholysis. Longitudinal erythronychia is more commonly found affecting digits of the hand than the foot. This may reflect a greater patient awareness of fingernail than toenail changes or the fact that toenails are thicker and so less altered by the disease likely to manifest as erythronychia.

Longitudinal erythronychia may present as a single or paired band in 1 nail or as multiple bands in many nails (Figure 1). As a single band it is likely to reflect a focal matrix disease. Multiple bands can represent multifocal inflammatory disease such as lichen planus or Darier disease. The width of isolated LE is usually less than 3 mm and seldom progresses substantially. Consequently, the nail remains largely intact. The appearance and characteristics of individual bands vary within a presentation of multiple LE. In diseases such as lichen planus, the abnormality may extend the full width of the nail matrix, destroy nail-plate production, and create the “winged nail” sign termed pterygium. Herein, we describe 8 representative cases of LE with different causes to convey the spectrum of presentation of this sign.

**Report of Cases**

**Case 1**

Patient 1 was the only patient in the study with coincident disease (oral lichen planus) that might have been relevant to the presenting condition (Table). She was a 66-year-old white woman who presented with an 8-year history of a single, expanding, longitudinal red streak in the left thumbnail extending to the matrix. She had a 13-year history of biopsy-proven oral lichen planus and no other skin, scalp, or nail disease. She had negative patch test results, which included a battery using dental mercurial compounds. Longitudinal excision to the proximal matrix revealed focal parakeratosis in the matrix and a prominent granular layer, multinucleated cells in the nail bed, and occasional koilocytes in the onychocorneal region.

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Healing resulted in a normal nail with some persistent fragility at the distal edge along the line of the original abnormality.

**CASE 2**

A 50-year-old white woman presented with an 8-month history of a progressive longitudinal red streak in the right thumbnail 2 mm in width extending into the matrix. The split was prone to catch and extend to the middle of the nail bed, which caused the patient pain. Because the change was in the lateral 5 mm of the nail unit, she opted for full excision of the lateral fraction of the nail unit. This was the only way to guarantee that there would be no future split. Histologic analysis revealed eosinophilic cells in stratified layers, a granular layer (which is usually absent in the normal nail bed), and areas of parakeratosis in the matrix and nail bed, but no specific disease.

**CASE 3**

A 36-year-old woman presented with a 3-year history of a fluctuating but nonprogressive red streak in the right thumbnail. At times, a small keratosis emerged at the distal edge in line with the streak and caused thumb pain, which negatively affected her work as a painter and decorator. Examination revealed a longitudinal red streak of 1 to 2 mm in width that extended into the matrix. At the distal edge was an irregularity aligned with the streak and a small yellow keratosis beneath the nail plate. Histologic analysis after excision of the streak to the proximal origin revealed nonspecific findings with hyperplasia but no dysplasia. Healing was uneventful, and a normal nail regrew.

**CASE 4**

A 46-year-old male farmer presented with an 8-month history of a nonprogressing longitudinal split in the left

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**Clinical Characteristics, Treatment, and Results**

<table>
<thead>
<tr>
<th>Patient No./Sex/Age,y</th>
<th>Duration of Disease, y</th>
<th>Affected Digit</th>
<th>Distal Keratosis</th>
<th>Excision</th>
<th>Result</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/F/66</td>
<td>8</td>
<td>Right thumb</td>
<td>Yes</td>
<td>Full length</td>
<td>Cure with fragile nail</td>
<td>Onychopapilloma</td>
</tr>
<tr>
<td>2/F/50</td>
<td>0.75</td>
<td>Right thumb</td>
<td>No</td>
<td>Lateral nail unit</td>
<td>Cure</td>
<td>Eosinophilic granules with parakeratosis</td>
</tr>
<tr>
<td>3/F/36</td>
<td>3</td>
<td>Right thumb</td>
<td>Yes</td>
<td>Full length</td>
<td>Cure</td>
<td>Hyperplasia, possibly viral</td>
</tr>
<tr>
<td>4/M/46</td>
<td>0.75</td>
<td>Left thumb</td>
<td>No</td>
<td>Matrix dermis</td>
<td>Cure</td>
<td>Prominent glomus structures</td>
</tr>
<tr>
<td>5/F/58</td>
<td>3</td>
<td>Right thumb</td>
<td>Yes</td>
<td>Distal nail bed</td>
<td>Recurrence</td>
<td>Onychopapilloma</td>
</tr>
<tr>
<td>6/M/75</td>
<td>2</td>
<td>Left thumb</td>
<td>Yes</td>
<td>Full length</td>
<td>Cure, with distal onycholysis</td>
<td>Onychopapilloma</td>
</tr>
<tr>
<td>7/M/70</td>
<td>2</td>
<td>Right thumb</td>
<td>Yes</td>
<td>Full length</td>
<td>Cure</td>
<td>In situ squamous cell carcinoma</td>
</tr>
<tr>
<td>8/M/57</td>
<td>4</td>
<td>Left third finger</td>
<td>No</td>
<td>Matrix dermis</td>
<td>Cure</td>
<td>Glomus tumor</td>
</tr>
</tbody>
</table>

A longitudinal erythronychia with hemorrhage distally. A keratosis is noted beneath the free edge (A) in line with the red streak (B). There is no split.
Clinically, a 1.5-mm-wide longitudinal red streak in the nail extended into the matrix. At the distal edge, a split extended up to 4 mm into the nail bed with no underlying keratosis. At surgery, the exposed matrix appeared to bulge at the origin of the streak. Treatment was therefore directed at excising the dermal tissue through a longitudinal incision in the matrix at this site. Histologic analysis revealed hyperplastic glomus structures but no tumor. The nail healed with no streak or split.

CASE 5

A 58-year-old woman presented with a 3-year history of a nonprogressive red streak in the right thumbnail representing a cosmetic handicap. Clinically, a 2-mm longitudinal red streak in the nail plate extended between the matrix and a distal V-shaped nick with associated keratosis. The patient did not want formal excision with concomitant risk of postoperative dystrophy but requested excision of the distal keratosis and nail bed. This was done without avulsion. There was temporary improvement, but the keratosis regrew within 4 months. There was no other change in the streak or nail. Histologic analysis revealed multinucleated giant cells, parakeratosis, and no dysplasia.

CASE 6

A 75-year-old man presented with a 2-year history of a dual LE of the left thumb. Two streaks, each 1.5 mm wide, extended from the matrix to the free edge. Both contained splinter hemorrhages in the distal 20%. A small keratosis could be found intermittently at the free edge beneath the nail. The abnormality was excised into the matrix. Long-term sequelae included a small field of distal onycholysis but no split. Histologic analysis revealed multinucleated giant cells, parakeratosis, and no dysplasia.

CASE 7

A 70-year-old man complained of a 3- to 4-mm red streak in the nail with minimal discomfort (Figure 3). The streak had enlarged over a 2-year period. Clinically, there was a slight split of the nail distally with a subungual keratosis and associated focal onycholysis. Excision revealed in situ squamous cell carcinoma within the matrix and proximal nail bed. Healing occurred with no split or recurrence over 24 months.

CASE 8

A 57-year-old man presented with the classic history of a glomus tumor of the left third finger causing pain particularly in the cold and at night. A small focal matrix discoloration with a red band extended distally to the free edge. At times, the free edge split, reflecting the thinned nail. There was no distal keratosis. Excision of the dermal matrix tumor through a slit in the matrix epidermis cured the symptoms and clinical signs.

COMMENT

These cases illustrate several important points concerning the clinical presentation, diagnosis, and treatment of localized LE. The problem most commonly presents in the thumb in middle age. The subjective problem may be the inconvenience of catching the fragile split’s free
edge, discomfort, or an issue of diagnostic or cosmetic concern. Any of these points may overlap.

Many of the surgical and therapeutic considerations are shared with those of a longitudinal melanonychia. A balance must be struck between diagnosis, therapy, and risks of the complications of intervention. Where the disease is definable to the matrix with a dermal tumor such as a glomus tumor, focal dermal excision through a slit in the matrix may result in a cure, as in cases 4 and 8. When distal keratosis is present, longitudinal excision may result in cure as long as it extends all the way back into involved matrix. Excision of the keratotic focus beneath the distal free edge nick does not result in cure, as illustrated by case 5. It also fails to yield accurate diagnostic material and would have represented a false-negative diagnosis in cases 4, 7, and 8. This could be predicted with the clinical origin of the erythronychia reflecting a matrix or submatrix location. However, it may not always be clear whether the keratosis is part of a primary disease or part of an LE. Examination with a dermatoscope can help reveal subtle continuity of the abnormality with the matrix.

A distal keratosis is seen in Darier disease where there are multiple bands of LE terminating with a V-shaped notch. In Darier disease, the primary abnormality is thought to be within the matrix. This observation supports attributing the distal keratosis to a primary matrix process, although the mechanism whereby this happens is not clear. Multinucleated cells are often seen in the distal nail bed in Darier disease, subungual warty dyskeratoma, and onychopapilloma, which indicates that it might be a nonspecific response. When there is a diagnosis of onychopapilloma, the presence of koilocytes raises the possibility of viral origin. Onychopapilloma is a clinical description of an LE with distal subungual keratosis with some clinical and histologic characteristics of a wart. However, while onychopapilloma may be a convenient term, there remains uncertainty whether it is a discrete entity and whether it is related to human papilloma virus infection.

Medical concern is legitimate in any lesion when an undiagnosed condition evolves and particularly when it is associated with destruction, such as a split in the nail. Magnetic resonance imaging may help define a focal

Figure 3. In situ squamous cell carcinoma presenting as longitudinal erythronychia.

Figure 4. The anatomic and pathologic basis of erythronychia.

Anatomic Basis
- Focal Loss of Function in Distal Matrix

Pathologic Basis
- Dermal Tumor; Pressure
  - Eg, Glomus Tumor
- Vascular Swelling
- Matrix Epidermis Disease
  - Eg, Darier Disease
- Dysplasia
- Lichenoid Disease
- Possible Viral Infection?

(a)

(b)

(c)

(d)

(e)

(f)
tumor and is particularly relevant when pain is present, suggesting a glomus tumor. Time may be a useful part of evaluation if diagnosis alone is the concern. Given how slowly nails grow, a 6- to 12-month period is usually necessary. In common with longitudinal melanonychia, when a band broadens, the passage of time may limit the options to successfully excise the disease and restrict any diagnostic biopsy to an incisional sample. Such a narrow biopsy might miss the diagnostic focus because there is no pigment to indicate where the most informative specimen might be taken. For this reason, we recommend that when surgical intervention is clinically warranted, diagnostic biopsies of LE should be avoided in favor of excisional biopsies. In further support of this recommendation, there is a higher chance of scarring after matrix biopsies if they are performed at a site of previous surgery.

Case 7 illustrates why surveillance and an open mind are important. Clinically, Bowen disease evolves to distinguish itself from the classic localized LE. A greater degree of onycholysis was seen in this case. But the early stages of a dysplastic process and multifocal inflammatory disease such as lichen planus can all manifest a single LE, as was seen in cases 1 through 5.

All of these processes have focal loss of matrix function as a common feature (Figure 4). Within the longitudinal axis, this results in a streak of thinned nail and creates a ventral groove. A thin nail is more transparent, and this enhances the perception of nail bed blood vessels. In addition, the normal nail at the margins compresses the nail bed, highlighting engorgement in the nail bed entrapped within the groove (Figure 5). This will dispose to hemorrhage, as reflected in the splinter hemorrhages seen in some cases. At the free edge, 4 of the present 8 cases had an irregularity or split. Where the nail is thinner, it is at its oldest and is subjected to the most trauma.

From the present series, it is apparent that localized LE warrants assessment and possibly histologic investigation. Dermatoscopy improves local assessment. Surgery may not always reveal a discrete pathologic process but when targeted to the clinical clues can be curative.

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REFERENCES