

Subungual Leiyomyoma Presenting as Erythronychia: Case Report and Review of the Literature

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ABSTRACT

We report a rare case of leiomyoma of the fingernail in a 59-year-old woman. She presented with red discoloration, lifting, and distal splitting of her left 2nd fingernail for several months. She reported sensitivity at baseline which became more painful in the cold. Histopathology sections from the nail matrix biopsy specimen showed a dermal proliferation of bland appearing spindle shaped cells with elongated, blunt ended nuclei (SMA and caldesmon positive), arranged in fascicles, which is typical of leiomyomas. Interestingly, our patient had a history of uterine leiomyoma, requiring hysterectomy. To our knowledge, this case is the first report in which a subungual leiomyoma is associated with another leiomyoma in the same patient.

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INTRODUCTION

Leiomyomas are benign neoplasms derived from smooth muscle and are primarily found in the gastrointestinal and female genital tract. In the skin, leiomyomas present as dermal-based nodules or papules with smooth borders and firm consistency. There are three main subtypes, which include: piloleiomyoma, arising from arrector pili muscles, genital leiomyoma, derived from smooth muscle of the scrotum, vulva, or nipple, and angioleiomyoma, arising from the vein smooth muscle.[1] Angioleiomyomas most commonly occur within the 30-50 year age range and are twice as prevalent in women than in men. They most frequently involve the lower extremities, with the upper extremities affected in <10% of cases. Malignant transformation is rare, and tenderness is a common symptom, although most are asymptomatic.[2]

CASE REPORT

A 59-year-old woman presented with red discoloration of her left 2nd fingernail for several months. She also noticed lifting of the nail and distal splitting that then progressed proximally. She reported that she had some sensitivity of the affected nail at baseline and it became somewhat painful in the cold. She denied prior trauma to the nail, bleeding, or purulent discharge. Her medical history was significant for Grave's disease, hyperlipidemia, and uterine fibroids requiring hysterectomy. Her only medication was methimazole. On physical examination, the left 2nd fingernail demonstrated longitudinal red discoloration near the lunula that blanched with pressure, as well as, severe onycholysis and longitudinal splitting (Figure 1). All other fingernails and toenails were unremarkable. The Love test and cold sensitivity test were negative. An x-ray of the affected nail unit was negative for bone erosions or tumors. An MRI with and without contrast revealed a 1 x 1 x 3 mm enhancing lesion with bony erosion, which was suspicious for a glomus tumor. A nodule underlying the area of erythronychia was exposed and biopsied following partial nail avulsion. The specimen was analyzed by histopathology (Figures 2-4).

FIGURE 1. Clinical appearance of the left 2nd fingernail. The left 2nd fingernail with longitudinal red discoloration near the lunula, severe onycholysis and longitudinal splitting.

Histologic sections of the biopsy specimen showed a dermal proliferation of bland appearing spindle shaped cells with elongated nuclei, arranged in small fascicles (Figures 2, 3). The spindle shaped cells were marked strongly and diffusely with caldesmon (Figure 4) and with smooth muscle actin (SMA) immunostains. S100 and CD31 immunostains were negative. Eight weeks following surgery, the nail sensitivity had resolved and the erythronychia was no longer present.

FIGURE 2. Histopathology of the biopsy specimen. A dermal based proliferation of spindle shaped cells with elongated nuclei, arranged in small fascicles (H and E section, 4x).

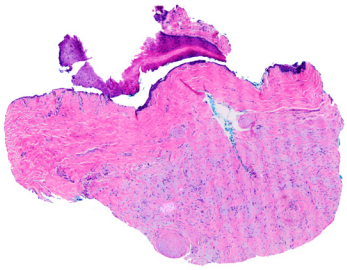


FIGURE 3. Histopathology of the biopsy specimen. A dermal based proliferation of spindle shaped cells with elongated nuclei, arranged in small fascicles (H and E section, 20x).

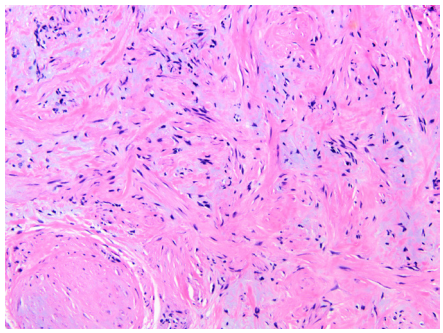
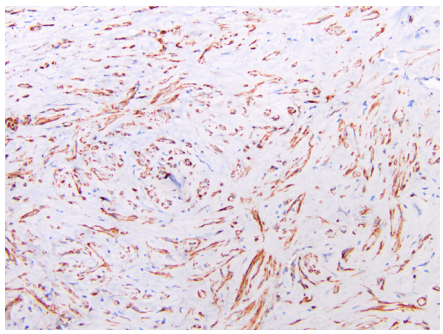


FIGURE 4. Histopathology with caldesmon immunostain. The spindle shaped cells are highlighted by the caldesmon immunostains.



DISCUSSION

Our patient presented with longitudinal erythronychia, onycholysis, and nail splitting along with cold sensitivity and an enhancing lesion with bony erosion on MRI. The differential diagnosis for monodactylous longitudinal erythronychia, defined as longitudinally oriented red line in the nail plate, includes onychopapilloma, lichenoid inflammation, glomus tumor, verruca, squamous cell carcinoma, and amelanotic melanoma.[3] Onychopapilloma is the most common cause of longitudinal erythronychia, which presents with a keratotic papule at the free edge of the nail plate, which was notably absent in this case. The Love test (pinpoint pressure provoking pain in the area of the tumor) and the cold sensitivity test (cold application reproduces pain), when positive are highly suggestive of a glomus tumor.[3] Despite the enhancing lesion on MRI with bony erosion suspicious for a glomus tumor, both the Love and cold sensitivity test were negative in this case.

Our patient's biopsy was consistent with a subungual leiomyoma. A leiomyoma, including a subungual leiomyoma, on microscopic examination exhibits a proliferation of bland appearing spindle shaped cells with elongated, blunt ended nuclei (SMA and caldesmon positive), arranged in fascicles. Histologically, an onychopapilloma represents a benign proliferation of nail bed epithelium characterized by acanthosis, papillomatosis, and matrix metaplasia. A glomus tumor is a modified smooth muscle cell neoplasm (SMA positive), which on microscopic examination appears as a distinct proliferation of bland appearing round cells (glomus cells), surrounding vascular spaces. Squamous cell carcinoma and melanoma are cytologically atypical and mitotically active proliferations of epithelial cells or keratinocytes (cytokeratin positive), and of melanocytes (S-100, melan-A and HMB45 positive), respectively. Verruca, on histologic examination, is a papillomatous growth of keratinocytes associated with viral inclusion bodies or koilocytes. A lichenoid inflammatory process presents as a band-like infiltrate which obscures the dermal-epithelial junction.

Leiyomyomas involving the nail unit or subungual leiomyomas are quite rare with the first case reported by Lebouc in 1889.[4] Subsequently, only 5 reported cases have been reported to date, including our case.[1,2,5,6] Of these cases, the mean age was 50 years (range 16-69), and there was a female predominance (4/5 patients). Three cases involved the index finger (2 left, 1 right), and the other cases involved the right hallux and left thumbnail. Duration of the nail change was several months in 3/5 cases, and the two other cases for 2 years and 10 years. The most common physical examination findings were longitudinal erythronychia or less commonly a blue nodule. Other secondary findings were onychoschezia and onycholysis. Symptoms included pain, tenderness and/or cold sensitivity. X-rays were typically negative for bone erosions or masses. MRI with contrast typically showed bone erosions and a well-defined tumor, and the vasculature in the case of angioleiyomyoma was enhanced on post-contrast images.[6]

Interestingly, our patient had a history of uterine leiyomyomas, requiring hysterectomy. To our knowledge this is the first report of the association of a subungual leiyomyoma with other leiyomyomas in the same patient and suggests a common pathogenic mechanism. While rare, leiyomyoma of the nail unit should be considered in the differential diagnosis of longitudinal erythronychia, particularly when accompanied by pain. A biopsy with histopathology is necessary for diagnosis, since glomus

tumors, as well as, nail unit squamous cell carcinoma and malignant melanoma can present similarly.[3]

DISCLOSURES

The authors have no conflicts of interest.

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