Linear atrophoderma of Moulin: A distinct entity?

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Abstract

Linear atrophoderma of Moulin (LAM), morphea, and atrophoderma of Pasini and Pierini (APP) show similar clinical and histological features. Despite theses similarities, some have classified them into distinct diseases. We present a case which has features of all three entities and supports the hypothesis that these diseases represent a spectrum rather than distinct diseases.

Our case is a16-year-old Caucasian male who presented with a six year history of asymptomatic, multiple brown areas located predominantly on his left side. His clinical presentation was consistent with a diagnosis of linear atrophoderma of Moulin (LAM). A biopsy was performed which showed alterations more consistent with morphea due to the presence of thickened collagen bundles. However, many case reports of LAM have described thickened collagen bundles present on histological examination. LAM is a rare, acquired linear atrophoderma that typically manifests in childhood or adolescence. In general, there is no preceding inflammation, induration, or resultant sclerosis. The depressed nature of the lesions is speculated to be due to a reduction in subcutaneous adipose. The clinical presentation and histological findings are also similar APP. Differentiation is made clinically by APP's lack of distribution along Blaschko's lines. APP is considered to be a type of superficial, abortive morphea in which sclerosis fails to develop. The clinical and histological similarities of APP and LAM raises the possibility that these diseases represent a spectrum of superficial morphea rather than distinct entities.

Case Presentation

➢**HPI:** 16-year-old Caucasian male presented with a six year history of asymptomatic, hyperpigmented depressions located primarily on his left side. The lesions began developing at 10 years of age and had completely developed by 13 years and have since remained stable. No erythema, induration, or violaceous border surrounding the lesions was reported by the patient or family members. A trial of an unknown topical medication, thought to be an antifungal by the patient's parents, resulted in no improvement. No further treatment attempts were made.

- ➢**PMH:** No current or past medical issues, normal birth and development.
- ➢**Family:** Noncontributory. One biological brother without any medical problems.
- ➢**Social:** (+) tobacco use (cigarette smoker)
- ➢**Medications:** No current medications
- ➢**Allergies:** NKDA

➢**PE:** Hyperpigmented, depressed, linear plaques along Blaschko's lines on the left upper extremity, axilla, left lateral chest wall extending anteriorly to the left areola, left posterior lower extremity, and right posterior shoulder. No erythema or induration was noted (Figure 1).

➢An incisional, elliptical biopsy was performed on the left lower extremity incorporating lesional and perilesional normal skin.

➢**Histopathology:** Microscopic examination revealed a normal epidermis, thickened collagen bundles in the mid dermis entrapping eccrine ducts, sparse perivascular lymphocytic infiltrate, and thickened hyalinized collagen in the deep dermis .Verhoeff-van Gieson (VVG) stain revealed thickened dermal elastic fibers compared with perilesional normal skin (Figure 2).

Results

Figure 1

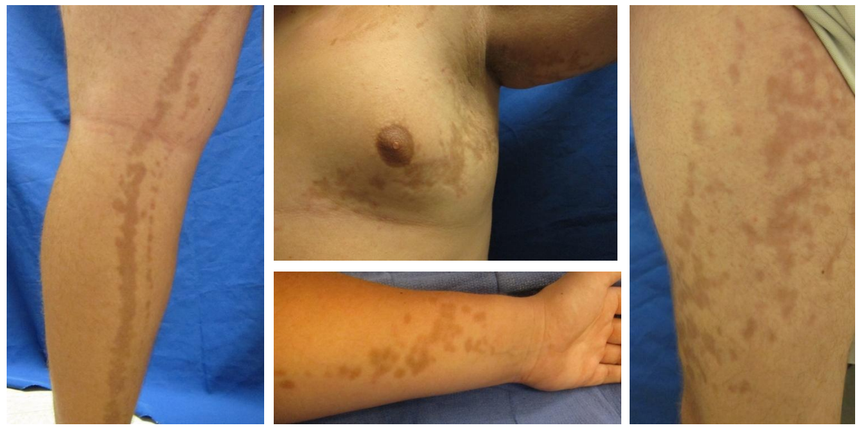
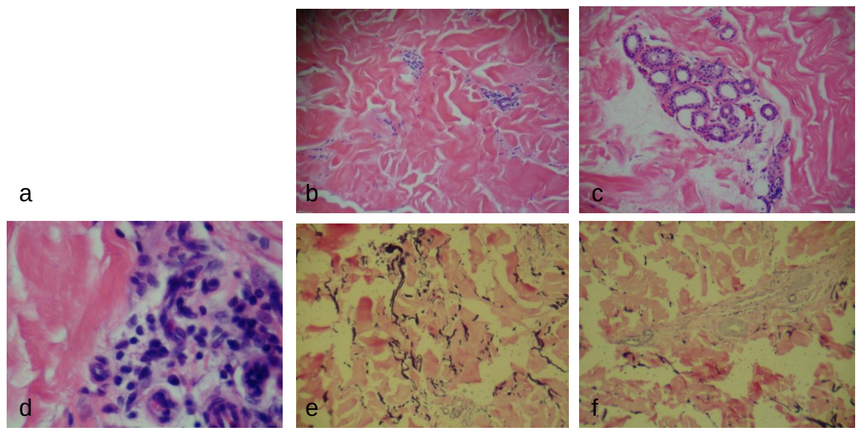


Figure 2



(a)H&E at 40x showing normal epidermis with thickened collagen bundles in dermis (b) thickened collagen in mid reticular dermis (c) collagen entrapping eccrine ducts (d) dermal lymphoplasmacytic infiltrate (e) VVG of lesional skin with dense, thickened dermal elastic fibers (f) VVG of normal perilesional skin

Figure 3

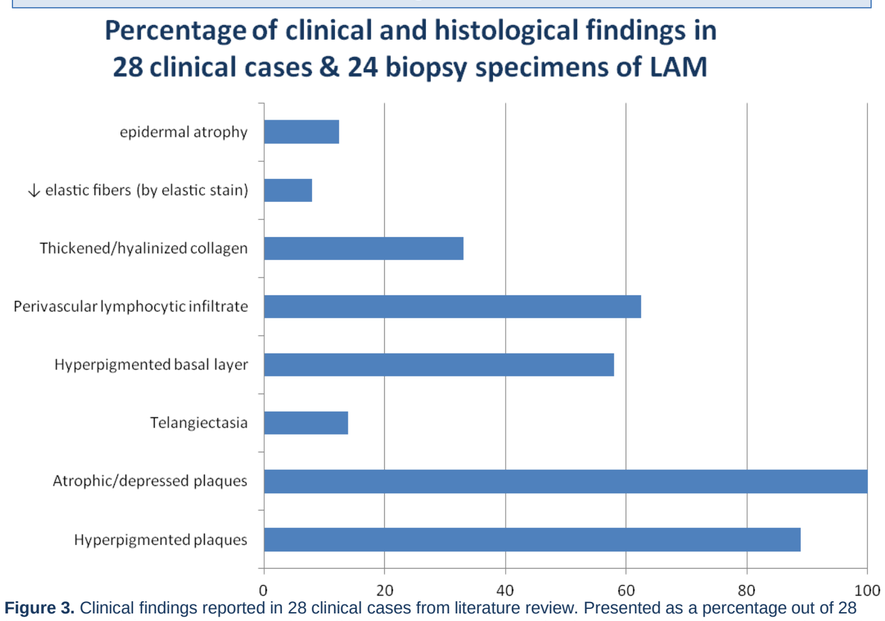


Figure 3. Clinical findings reported in 28 clinical cases from literature review. Presented as a percentage out of 28 total cases. Histological findings reported in 24 biopsy specimens from literature review reported as percentage out of 24. (References 1-20)

Discussion

Linear atrophoderma of Moulin (LAM) is a rare, acquired, atrophic band like skin lesion that often shows:

- ➢ Hyperpigmentation (25/28), occasional telangiectasias (4/28), and always follows the lines of Blaschko.
- ➢Age of onset ranges from < 1 year to 37 years (mean age, 14.9 yrs).
- ➢Usually asymptomatic lesions(26/28) and stabilize after a period of rapid progression.
- ➢Histologically, the atrophic lesions demonstrate variability in epidermal and dermal changes including hyperpigmentation of the basal epidermis (14/24),dermal perivascular lymphocytic infiltrate (15/24), thickened or hyalinized dermal collagen (8/24), acanthosis (6/24), epidermal atrophy (3/24), and decreased or fragmented elastic fibers demonstrated by stain for elastic fibers (2/24).

Clinically and histologically, LAM is similar in presentation to atrophoderma of Pasini and Pierini (APP):

- ➢ APP similarly shows atrophic patches with frequent hyperpigmentation . Histologic findings of APP are variable and include hyperpigmentation of the basal epidermis, mild perivascular mononuclear cell infiltrate, clumping of collagen fibers, and normal to clumped or fragmented elastic fibers.
- ➢APP is differentiated from LAM chiefly by its lack of distribution along Blaschko's lines.
  - ❖The Blaschkolinear distribution of LAM is thought to be due to a genetic mosaicism as is seen in other Blaschkolinear diseases (i.e. segmental Darier's disease and CHILD syndrome).
  - ❖ The primary process of LAM, however, is most likely independent of this mosaicism contributing to its distribution along Blaschko's lines.
  - ❖Therefore, the primary process may represent the same etiology as APP, given the clinical and histological similarities.

APP, morphea, and LAM may thus represent a spectrum rather than three distinct disease entities:

- ➢APP is considered by many to be a an abortive morphea in which induration fails to develop. The similar clinical and histologic manifestations of APP and LAM indicate that if APP is truly a variant of abortive morphea, LAM most likely also belongs on the morphea spectrum.
- ➢The presence of a perivascular lymphocytic infiltrate in 62.5% of LAM cases as well as the presence of dermal collagen changes in 33% creates further overlap between these three diseases.
- ➢Due to the overlap in clinical and histological presentation between morphea, APP, and LAM, we postulate these diseases most likely represent a spectrum of disease rather than distinct entities.

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