Acrosyringeal Nevus in a Patient with ILVEN

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26 y.o. woman initially presented for evaluation of multiple cutaneous lesions on her chest, left upper and lower extremity, and genital region that had been present since birth.

These sites have intermittently flared in the past, and were currently associated with significant pruritus and pain.

Denied any other associated symptoms, and ROS otherwise unremarkable.

The patient was previously treated with multiple laser and surgical procedures on the trunk and left upper extremity 10-15 years prior with moderate improvement in cosmesis.
Prior Treatment
Prior Treatment
Prior Treatment
Physical Examination

- Multiple, coalescing erythematous to brown verrucoid patches and plaques with areas of scar formation were noted on the chest, left upper and lower extremity, left buttocks, and genital region.
Physical Examination
Physical Examination
Physical Examination
Physical Examination
Physical Examination
Impression

Given the patient’s history and physical examination findings, a presumptive clinical diagnosis of Inflammatory Linear Verrucous Epidermal Nevus Syndrome (ILVEN) was made.
Course and Therapy

- Combination therapy of topical fluocinonide and tacrolimus ointment, and IL TAC at 10 mg/ml was initiated.

- At follow-up, the patient noted a significant symptomatic improvement of her condition.

- However, she did complain of a nodule on the left heel that had become progressively tender.

- Lesion had been present for several years, but had gradually increased in size and became exceedingly painful.
Physical Examination

- An approximate 1.8 X 1.5 cm indurated, erythematous to brown plaque (studded with a nodule) and minimal overlying scale was noted on the left heel.
Left Heel Skin Lesion
Left Heel Skin Lesion
The patient elected surgical removal of the neoplasm.

An elliptical excision was performed to provide the patient with the best functional and cosmetic outcome.

Specimen was sent out for histopathologic examination.

Upon subsequent follow-up visit, the patient noted a significant functional and symptomatic improvement of her surgical site.
Histopathology

- Proliferation of acrosyringeal keratinocytes with focal areas of PAS positivity extending into the dermis as thin anastomosing cords of cells from the base of an acanthotic epidermis.

- Associated dermal fibrosis with thickened collagen and increased fibrocytes and scattered plasma cells were also noted.
Histopathology
Case Discussion

- ILVEN: rare genetic dyskeratotic disease characterized by pruritic, erythematous scaly lesions that form as linear bands following the lines of Blaschko.

- Likely caused by a lethal AD mutation that is rescued by genetic mosaicism.

- Commonly affects children with skin lesions progressing into puberty, with females more often affected than males.
ILVEN: unknown pathogenesis; however upregulation of IL-1, IL-6, TNF-alpha, and ICAM-1 may be involved.

Association of ILVEN with arthritis and autoimmune lymphocytic thyroiditis from previous cases.
Case Discussion

- Clinical presentation: ILVEN displays inflammation that may intensify and is marked by verrucous lesions grouped together in a linear distribution.
- Lesions may be erythematous and pruritic, causing moderate to intense discomfort.
Case Discussion

- Histology: ILVEN is characterized by elongation of the epidermal rete ridges with areas of hypergranulosis and orthokeratosis alternating sharply with regions of parakeratosis without an underlying granular layer.
Alternating blue and red corneum

Zones of loss of granular layer
Case Discussion

- Treatment: difficult and transient with ILVEN.
- Symptomatic relief with topical corticosteroids and calcipotriols.
- Systemic treatment with acitretin and etanercept.
- Laser ablation, electrofulguration, cryotherapy, and medium to full-depth chemical peels for smaller lesions.
- Full-thickness surgical excision.
Case Discussion

- Ilven has been associated with several disease processes.
- No apparent correlation with acrosyringeal nevi.
Acrosyringeal Nevus

- Rare, benign neoplasm first reported by Weedon and Lewis as a solitary lesion on the dorsal hand.

- Consists of a proliferation of PAS-positive acrosyringeal keratinocytes extending down from the undersurface of the epidermis as thin, anastomosing cords into the dermis.

- Abundance of stromal plasma cells is also characteristic.
Acrosyringeal Nevus

- Lesions may appear linear, plaque-like, or multiple in nature.
- Diffuse lesions noted in ectodermal dysplasia.
- Distal extremities appear to be favored location.
- While considered benign in nature, lesions may be removed due to diagnostic uncertainty, cosmetic improvement, or symptomatic relief.
Acrosyringeal Nevus

- The unique histopathological features of these lesions allow for distinction from eccrine syringofibroadenomas.
  - Lack PAS-positivity and stromal plasma cell proliferation.

- Typically also benign but cases of malignant transformation reported.

- To date, no apparent malignant potential associated with acrosyringeal nevi.
Acrosyringeal Nevi

- Eccrine poromas share similar histologic features.

- Lesions may be differentiated by thinner anastomosing cords of cells and bud-like projections of tumor present at intervals along the basal layer seen in acrosyringeal nevi.

- Eccrine poromas may also display cystic spaces within the tumor or stromal telangectasias.
Acrosyringeal Nevi

- Clinical case of an acrosyringeal nevus in a patient with ILVEN is the first apparent case in the literature.

- Highlights the importance of a thorough evaluation for proper diagnosis and management of these patients.


References

