ZOSTERIFORM LICHEN PLANUS: AN UNUSUAL CLINICAL VARIANT

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AOCD MIDYEAR MEETING
APRIL 23-26 2015
No Financial Relations with Commercial Interests
OBJECTIVES

- Case presentation
- Introduction
- Epidemiology
- Clinical
- Pathogenesis
- Differential Diagnosis
- Histopathology
- Treatment and Prognosis
CASE PRESENTATION

• 52-year-old woman w/ six-week history of a pruritic eruption on her right leg

• PMHX: Discoid Lupus Erythematosus (DLE). No prior hx of herpes zoster

• No significant family or medication history

• Hepatitis C serology was negative
PHYSICAL EXAMINATION

- Purple pruritic papules and plaques with an overlying white scale present on her right thigh

- The lesions were arranged in a linear pattern in the L1, L2, and L3 dermatomal distribution

- A lacy white patch was seen on her left buccal mucosa

- There was no scalp or nail involvement
DIAGNOSIS

• Histopathology: Wedge shaped hypergranulosis, acanthosis, saw-tooth rete ridges, and a lichenoid infiltrate

• A diagnosis of Zosteriform Lichen Planus was made based on the clinical and pathological correlation
TREATMENT

• Intramuscular injection of Kenalog 40mg

• Tacrolimus ointment 0.1%, which was applied to the cutaneous and oral lesions twice daily
INTRODUCTION

• Lichen Planus (LP): Papulosquamous skin disorder with several morphological variants.

• Zosteriform LP may arise:
  - De novo
  - At sites of trauma (koebnerization)
  - Wolf’s isotopic response at the site of healed zoster

• Zosteriform type is an uncommon variant of lichen planus with dermatomal or zonal distribution.
EPIDEMIOLOGY

- Prevalence of LP is 0.22% to 5% worldwide
- Average age of onset is 50 years old
- No racial predilection
- Females slightly more affected often than males
- The mucous membranes are affected in 65% of cases
- Oral LP: Wickham's striae may develop into squamous cell carcinoma (SCC) in 0.2% of cases
- Zosteriform LP is an atypical presentation of linear LP. Linear LP accounts for less than 1% of cases
• Purple polygonal pruritic flat-topped papule and plaques. May have wickham's striae on papules

• Over 20 variants of lichen planus

• Linear LP refers to lichen planus with a unilateral linear distribution

• Can present in segmental distribution corresponding to a dermatome referred to as zosteriform lichen planus

• Zosteriform lichen planus:
  ▪ Wolf’s isotopic response at areas of healed zoster
  ▪ Secondary to Koebnerization from trauma
  ▪ De novo eruption on previously normal skin
Lesions arranged in band several centimeters wide and run along the course of a peripheral cutaneous nerve and its branches

- Triggered by neural factors
- Suggested that the lesions in zosteriform LP actually follow the Lines of Blaschko rather than an actual dermatome
- Blaschkoid lines are invisible lines in the skin that are believed to trace the migration of embryonic cells
- Some believe that true zosteriform LP only occurs if lesions develop at sites of healed herpes zoster
DIFFERENTIAL DIAGNOSIS

- Linear psoriasis
- Lichen striatus
- Linear epidermal nevus
- Linear darier’s disease
- Inflammtory linear verrucous epidermal nevus (ILVEN)
- Lichen simplex chronicus
- Lichenoid drug eruption
- Lichenoid mycosis fungoides
HISTOPATHOLOGY

- Band-like lymphohistiocytic infiltrate at the dermal-epidermal junction (DEJ) and upper dermis. Wedge-shaped hypergranulosis and acanthosis with saw-toothed rete ridges
  - Wickham’s Striae seen in areas of hypergranulosis

- Max Joseph Spaces: Vacuolar degeneration at the basal layer leading to focal subepidermal clefts

- Civatte Bodies: Eosinophilic remnants of anucleate apoptotic basal cells found in the dermis

- Squamatization: Maturation and flattening basal layer cells
DIRECT IMMUNOFUORESCENCE

• Direct Immunofluorescence (DIF):
  
  ▪ Shaggy fibrin, cytoid bodies, and deposition of IgM immunoglobulins at the DEJ
  
  ▪ Can distinguish LP from hypertrophic lupus erythematosus (continuous granular band of IgG, IgM, IgA, and C3 at the DEJ on DIF)
TREATMENT AND PROGNOSIS

• Treatments aimed to induce remission and relieve associated pruritus

• First line therapy:
  ▪ High potency topical steroids

• Systemic corticosteroids can be used as a second line treatment or in those with more extensive disease
TREATMENT AND PROGNOSIS

- Alternative treatments include

  - Immunosuppressive agents
    - Cyclosporine
    - Methotrexate
    - Azathioprine
    - Dapsone
    - Topical Tacrolimus

  - Narrow-band Ultraviolet B phototherapy. Psoralen plus ultraviolet A (PUVA)

  - Enoxaparin Sodium

  - Oral Metronidazole
TREATMENT AND PROGNOSIS

• Treatment of pruritus
  • Oral antihistamines
    • Diphenhydramine
    • Hydroxyzine
  • Topical antipruritic agents
    • Menthol
    • Camphor
    • Pramoxine
    • Doxepin

• Prognosis:
  • Often resolves over average period of 18 months
  • Approximately 20% of patients will have a second occurrence. In a subset of patients, the disease may persist for many years.

• Oral LP more therapy resistant, and close follow up advised given increased risk for SCC