Case 113

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Clinical Presentation

- 37 year-old male presented with 2 papules (6-8 mm)
  - Right shoulder
  - Right flank
- The lesions had been present for one month and were asymptomatic
Clonality Studies

- PCR for the T-cell receptor gene rearrangement (BIOMED2)
  - Positive for monoclonal T-cell population in both the right shoulder and abdomen lesions

- PCR for immunoglobulin heavy chain and kappa light chain gene rearrangement
  - Negative
Additional clinical history

• After ancillary studies were performed, we inquired about a detailed clinical history
  – The patient was found to have a history of seizure disorder and was taking phenytoin
Clinical Follow-up

• Phenytoin was discontinued with rapid and complete resolution of both lesions
• The patient has been well and without recurrence for 54 months
  – His seizures are now controlled by lamotrigine
Diagnosis

• Cutaneous atypical lymphoid hyperplasia ("pseudolymphoma") secondary to anticonvulsant (phenytoin) therapy

• Or as the panel suggested:
  – Lymphomatoid drug eruption
Monoclonal T-cell CLH

• Characterized most commonly by solitary lesions often of the head and neck
• Two histologic patterns described:
  – Nodular/diffuse
  – Superficial band-like
    • Common in drug associated cases
• Reactive entity characterized by an indolent clinical course and spontaneous regression
  – Drug associated cases: cessation often leads to regression

Case 136

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Clinical Presentation

• 35 year-old male with a solitary, painless erythematous plaque in the right axilla
  – Present for approximately one month and has increased in size
• He denies trauma to this area, systemic symptoms, or changes in his medications
Clonality Studies

• PCR for the T-cell receptor gene rearrangement (BIOMED2)
  – Positive for monoclonal T-cell population

• PCR for immunoglobulin heavy chain and kappa light chain gene rearrangement
  – Negative
Diagnosis

• Primary cutaneous CD4-positive small/medium T-cell lymphoma [SMPTCL] (WHO classification, provisional entity)
SMPTCL

- Characterized most commonly by solitary lesions often of the head and neck
- Low grade lymphoma with a dense nodular or diffuse dermal infiltrate of small to medium-sized monoclonal pleomorphic lymphocytes
- Characterized by an indolent clinical course with local therapy
- A follicular helper T-cell (T\textsuperscript{FH}) immunophenotype has recently been described in this entity:
  - PD-1+/CXCL13+/BCL6+

SMPTCL vs. T-cell CLH

• Studies for TCR monoclonality will help eliminate cases of non-clonal T-cell CLH from consideration

• Histologic clue:
  – Superficial band-like infiltrate is often seen in drug-associated Monoclonal T-cell CLH but is unusual in SMPTCL

• Significant morphologic overlap in this DDx:
  – PD-1 and other T\(^{FH}\) markers?
    • In our cases and in a recent study, these markers were expressed by atypical cells in both entities

SMPTCL vs. Monoclonal T-cell CLH

• Knowledge of clinical history is imperative in this DDx

• It is reasonable to express diagnostic uncertainty when a complete history is unavailable
Every life deserves world class care.