Dermatopathology
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DERMATOPATHOLOGY CASE CHALLENGE: RECOGNIZING MIMIS AND MASQUERADERS
Rosalie Elenitsas. University of Pennsylvania
Cases reported with histological features characteristic of the two entities in different areas of the subcutaneous tissue

Some cases respond to antimalarials, some progress to frank lymphoma

Some cases in between (indeterminate lymphocytic lobular panniculitis)
• 3 hipotesis
  • SPTCL con histological features mimicking those of LEP
  • LEP con features mimicking those of SPTCL
  • LEP y SPTCL were at the end of a spectrum of diseases. This would be supported by its frequent association
Plasmacytoid dendritic cells CD123+ in lupus erythematosus

- Clusters of plasmacytoid dendritic cells arranged close to the epidermis and in the subcutaneous tissue

- Characteristic finding of lupus that may be helpful for diagnosis but it is not specific (present in several other diseases)
ADVANCED DERMATOPATHOLOGY
Diagnostic criteria for skin involvement by IgG4-RD
(Lawrence Gibson. Mayo Clinic)

- One or two histologic features of dense lymphoplasmacytic infiltrate, fibrosis, and obliterative venulitis plus IgG4 plasma cell density more than 200/high-power field and IgG4/IgG plasma cell ratio more than 40%
- Serum IgG4 more than 135 mg/dL
- Other organ involvement

1: Probable diagnosis
1+2+3: Definitive diagnosis

Granuloma Faciale and Erythema Elevatum Diutinum in Relation to Immunoglobulin G4-Related Disease. An Appraisal of 32 Cases.


- Gibson et al reviewed 32 cases of GF or EED from Mayo Clinic to elucidate if they meet consensus histopathologic diagnostic criteria for IgG4-related disease
- All cases fulfilled presence of small vessel vasculitis, dermal fibrosis, and plasma cell infiltrates. Eosinophilic inflammation frequent
- Results:
  - No patient in this study fulfills consensus criteria for IgG4-RD (IgG4 plasma cell density more than 200/HPF and IgG4+/IgG+ plasma cell ratio>40%)
  - No patients have had systemic disease to suggest IgG4-related disease
- Conclusion
  - Despite some histopathologic, immunophenotypic, and clinical similarities between GF/EED and IgG4-RD, the cases studied do not meet the consensus diagnostic criteria to confirm inclusion of these skin disorders in the category of IgG4-RD
IgG4-related disease and the skin

• Skin lesions: papules, plaques or nodules in the head and neck area
• Most often patients have signs of disease elsewhere, especially in the glands of the head and neck area
• Elevated serum IgG4 important to diagnosis but not specific
• Allergic type signs and symptoms often present
• Skin biopsy should demonstrate elevated IgG4-bearing plasma cells; fibrosis may not be present
Pseudomyogenic hemangioendothelioma
(Omar Sangüeza. Winston Salem)

• Hemangioendothelioma sarcoma epithelioid-like

• 50 cases

• Clinical features: young adults, males>females (5:1), 20-50 years of age, most commonly lower extremities (other locs reported)

• Multifocal presentation

• More than 50% local recurrence, one patient metastasis to lymph nodes, one patient disseminated metastasis and died

Pseudomyogenic hemangioendothelioma

• Histopathological features

  • Poorly circumscribed neoplasm, involving dermis and/or subcutaneous tissue
  
  • Fascicular pattern with myxoid areas and occasional neutrophils
  
  • Large cells with vesiculous nuclei, prominent nucleoli, ample eosinophilic cytoplasm resembling rhabdomyoblats
  
  • Occasionally intravascular invasion
Pseudomyogenic hemangioendothelioma

• Differential diagnosis:
  • Epithelioid sarcoma
  • Epithelioid hemangioendothelioma
  • Epithelioid angiosarcoma

• Immunohistochemistry
  • Positive: AE1/AE3, CD31, ERG, FLI 1, INI 1, Vimentin
  • Negative: CD34, FXIIa, Podoplanin, Desmin, Myogenin, Myo D1, S100, CD10, CD99
FOSB is a Useful Diagnostic Marker for Pseudomyogenic Hemangioendothelioma Hung, Fletcher, Hornick. Am J Surg Pathol 2016

• A SERPINE1-FOSB fusion has been identified as a consistent genetic alteration in pseudomyogenic hemangioendothelioma

• Whole-tissue sections from 274 cases including 50 pseudomyogenic hemangioendotheliomas, 84 other vascular tumors and 140 other histologic mimics were evaluated

• Diffuse nuclear immunoreactivity for FOSB (>50% of cells) was observed in 48 of 50 (96%) pseudomyogenic hemangioendotheliomas