



Immunobullous Diseases: Paraneoplastic Pemphigus (PNP)

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Presenter:

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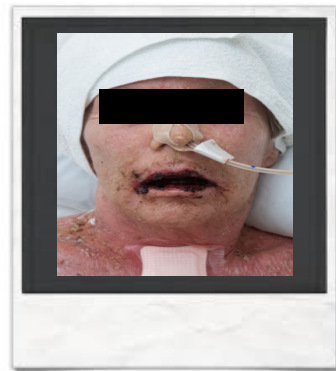
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Disclosures

- None

Paraneoplastic Pemphigus (PNP) aka Paraneoplastic Autoimmune Multiorgan Syndrome (PAMS)

- Rare, serious autoimmune blistering disease associated with an underlying malignancy
- Erosive changes of the oral mucosa and polymorphous skin lesions



Associated malignancies:

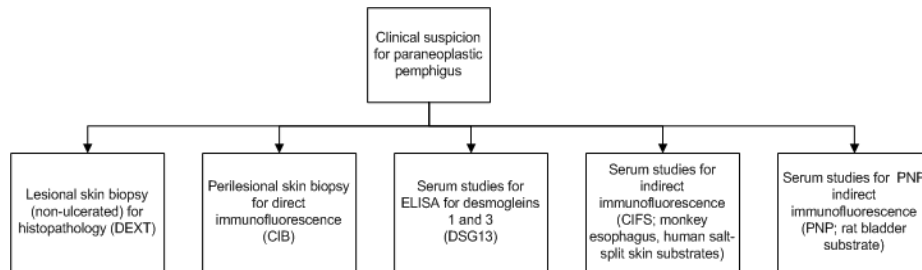
- Adults: Non-Hodgkin lymphoma, chronic lymphocytic leukemia, Hodgkin lymphoma, others
- Children: Castleman disease

Clinical complications of PNP/PAMS:

- Malignancy-related
- Infection
- Bronchiolitis obliterans

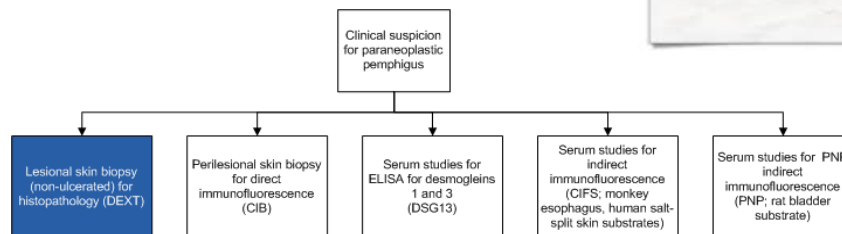
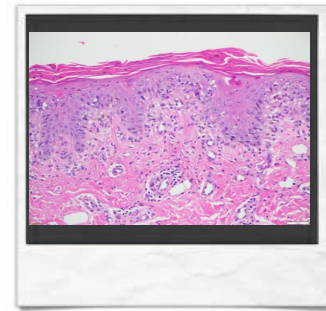
Laboratory Detection of PNP/PAMS

- Tissue-based and serum-based testing



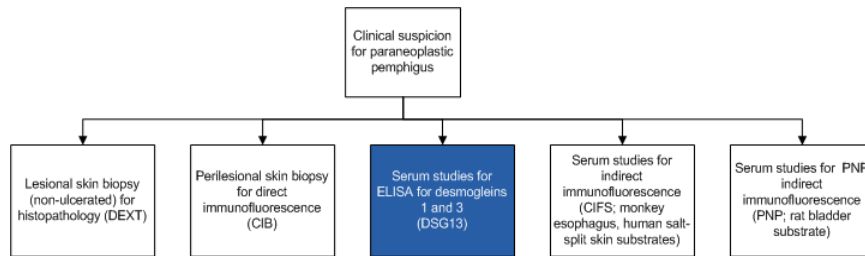
Histopathologic Findings:

- Acantholysis (66%)
- Keratinocyte necrosis (45%)
- Lichenoid infiltrate (45%)
- Subepidermal separation (23%)



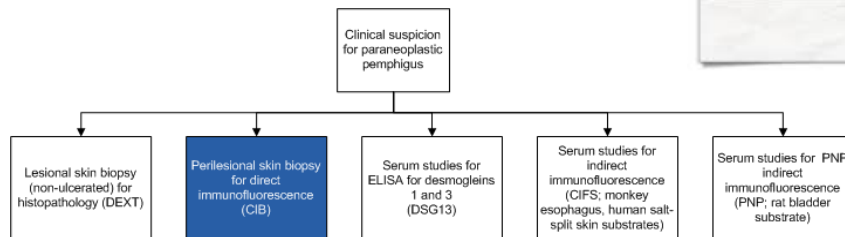
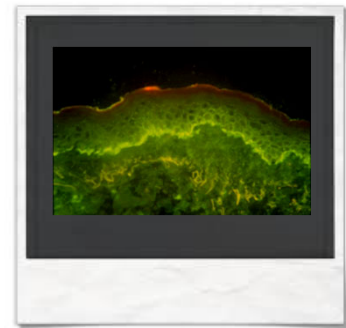
ELISA

- Sensitivity:
 - Desmoglein 1: 40%
 - Desmoglein 3: 71%



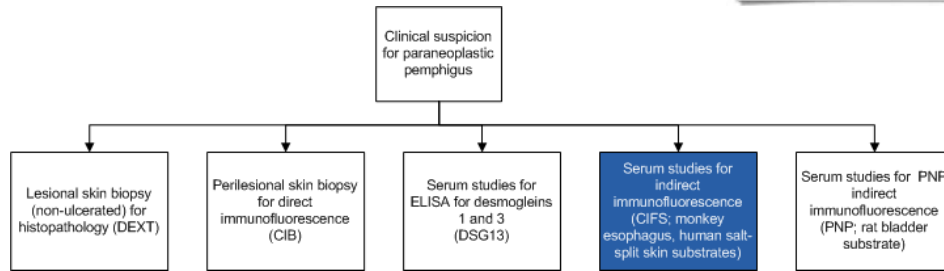
Direct Immunofluorescence Findings:

- Intercellular deposition of IgG and/or C3 (79%)
- Linear deposition (31%)



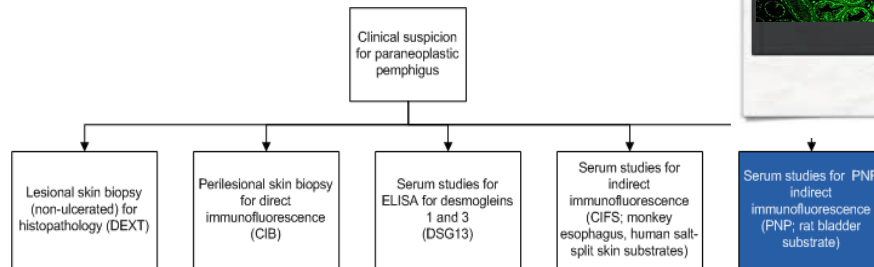
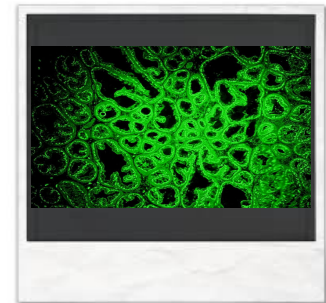
Indirect Immunofluorescence Findings:

- Intercellular deposition (75%)
- Intercellular and linear

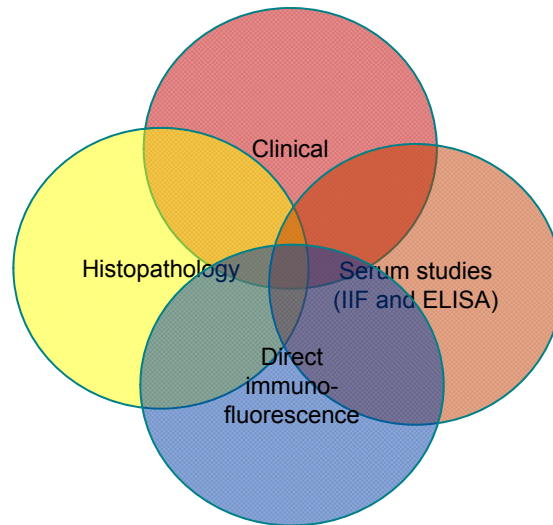


Indirect Immunofluorescence Findings (Rat Bladder):

- Intercell



Clinico-immuno-pathologic correlation



Hot Topics - PNP

- New Immunobullous Disease-Related Hot Topics coming soon!



Thank You