Case 1: ulcerative stomatitis

Dr Gilles F.H. Diercks
Center for skin blistering diseases
University Medical Center Groningen
The Netherlands
Female 37 years old

• Medical history:
  • For several weeks pain and ulcerations in her mouth
  • No blisters
Differential diagnosis (ulcerative) stomatitis

- Infection (candida)
- Lichen planus
- Chronic ulcerative stomatitis
- Lichenoid mucositis: medication, idiopathic
- Lupus erythematosus
- Autoimmune bullous diseases: pemphigoid, pemphigus vulgaris, paraneoplastic pemphigus
Further examination

- Painful fissures and erosions predominantly on hands and feet.
- 10 kilograms weight loss
Monkey esophagus  Salt-split skin
Immunoprecipitation

EP
PP
A2ML1
Criteria paraneoplastic pemphigus

• Major criteria
  • Severe stomatitis
  • Antibodies against envoplakin, periplakin and/or A2ML1
    • Immunoprecipitation  
    • Immunoblot and rat bladder  
  • Neoplasm

• Minor criteria
  • Physical examination: blisters, lichen planus-like lesions
  • Histology: suprabasal acantholysis or lichenoid dermatitis
  • Direct IF: ECS deposition or linear BMZ deposition of IgG +/- C3
  • Positive rat bladder
  • Antibodies against DSG1 and DSG3
  • Antibodies against BP230, desmoplakin

Follow-up

- Treatment with prednisolone and rituximab
- Perioperative human intravenous immunoglobulin

- After several weeks progressive dyspnoea: bronchiolitis obliterans
- (re)start rituximab and HIVIG: no effect
- Diseased under the diagnosis of bronchiolitis obliterans caused by paraneoplastic pemphigus
Paraneoplastic pemphigus

• Rare disease: UMCG 24 cases since 1997
• Associated with predominantly hematologic diseases
  • Castleman’s disease
  • non-Hodgkin lymphomas
  • dendritic cell sarcoma
  • thymoma
• Multiple antigens are involved. Most important ones: envoplakin, perilplakin and a2ml1
Take home messages

• Ulcerative stomatitis and an interface dermatitis could point out to \textit{paraneoplastic pemphigus}.

• Serologic investigations should be done by a specialized lab: immunoblot, immunoprecipitation and rat bladder.

• When positive search for an underlying neoplasm.

• Bronchiolitis obliterans is an important cause of dyspnoea and potentially lethal.