#AAD2019

Highlights AEDV

In 77TH AAD Congress

1-5 March 2019

★ Washington ★

Scientific Initiative of:

Sponsored by:

NOVARTIS

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Cutaneus manifestations in systemic disease

Dra. Águeda Pulpillo Ruiz

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ORAL SIGNS OF SYSTEMIC DISEASE: AUTOIMMUNE DISEASES

- **Behcet´s disease**

  Pathogenesis immune disregulation Th17 response

  Severity and mortality highest in yonguer.

  Cardioascular manifestations: arterial aneurysms pericarditis, endocarditis, intracardiac thrombosis

  Gastrointestinal manifestation dificult to distinguish between IBD

  AntiTNFα gastrointestinal ulcers
- **Cicatricial pemphigoid:** mycophenolate mofetil
- **Oral liquen plano** IVIG
- **Sjögren´s Syndrome**: minor salivary gland are submucosal biopsy excisional

ORAL SIGNS OF SYSTEMIC DISEASE: DRAMATIC ORAL DISEASES

Orofacial granulomatosis

- Melkersson-Rosenthal syndrome
- Miescher's cheilitis granulomatosa Monosymptomatic MRS Oligosymptomatic MRS
- Evaluation: Biopsy of oral lesión
- Exclude Crohn disease, sarcoidosis,
- Dental evaluation
- Patch testing (cinnamaldehyde, food coloring)

Pyostomatitis vegetans: IBD

Complex aphthosis

DD: RAS; trauma, intraoral herpes simplex, cyclic neutropenia

Drugs: Nonsteroidd antiinflamatory therapia , angiotensin converting enzyme inhibitors

Gluten enteropaty

Sodium lauril sulfate containing toothpaste

Helicobacter pylori

Laboratory: hematinic deficiencies: iron, B12, folic acid

UPDATES IN NEUTROPHILIC AND PUSTULAR DERMATOSES

Neutrophilic Urticaria

Urticarial rash wiht neutrophilic infiltrate perivascular, intersticial and often perieccrine tropism, leucocitoclasis without fibrinoid necrosis.

Associated diseases:

- Schitzler síndrome
- Lupus erytematosus
- CAPS: familial cold autoinflamatory síndrome; Muckle Wells Syndrome; cronic infantile neurologic cutaneous joints (CINCA). Mutacion NLPR3
- Adult onset Still´s disease
- Serum sickness-likes drug erupción.

Management:

Colchicina or dapsone

Schitzler CAPS: IL1 antagonist: anakinra, rilonacep, canakinumab

Still: Anakinra tocilizumab

UPDATES IN NEUTROPHILIC AND PUSTULAR DERMATOSES: UPDATES IN SWEET SYNDROME

Classic 50%; Malignant 30%; Drug induced 20%

Pathogenesis:

- Neutrophilic discrasia
- Diferentiation of malignan clones into mature neutrophis: drug induced SS all trans retinoic acid; fms-like tyrosin kinase-3 (FLT3)

Clinical

- Classic
- Pustular and bullous
- Necrotizing
- ND dorsal hands
- Giant cellulitis- like

Histopathologic

- Neutrophilic
- Histiocitoid
- Lymphocitic
- Subcutaneous
- Cryptococcoid

SS in pediatric population: consider interferopathia: CANDLE síndrome

UPDATES IN NEUTROPHILIC AND PUSTULAR DERMATOSES: UPDATES IN PYODERMA GANGRENOSUM

- **Associated comorbidities (67%)**
  Patients >65 yrs: -pathergy(36% vs 24%, p=.02) -RA, AS, malignancy,
  IBD more common in <65 yrs(47.7% vs 26.6%; p<.001)

- **Clinical subtypes**: Ulcerative PG; Bullous PG; Vegetative PG; Pustular PG; Peristomal

- **Drugs associated with PG** • cocaine, levamisole • isotretinoin, alitretinoin • PTU • adalimumab, etanercept, infliximab • azacitidine • gefitinib/ imatinib/ sunitinib • ipilimumab • enoxaparin • EPO, G-CSF • IFN

- **Treatment**
  Anakinra, canakinumab
  IL12/23. Ustekinumab
  IL17 :ixekizumab, brodalumab
  Apremilast (Laird et al, 2017, JAAD Case reports)
  Tocilizumab (Lee et al, 2016, JEADV)*
  JAK inhibitors:
    Tofacitinib (JAK1/3) (Kocharet al, 2019, Clin GE & Hep)
    Ruxolitinib(JAK2) (Nasifogluet al, 2018, BJD)