AEDV HIGHLIGHTS
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Systemic diseases

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Incidence of SLE & CLE is similar

New insights in LE pathogenesis ➔ New Treatments
- UV light ➔ Apoptotic KC ➔ IFN pathway ➔ Type I cytokines

Classic treatments:
- Potent topical corticosteroids
- Hydroxychloroquine (inhibit TLR7 & TLR9), Systemic Corticosteroids
- Immunosuppressants: MTX, Azathioprine, Mycophenolate
- Thalidomide: Inhibition of IFN gamma. side effects (teratogenicity, neurotoxicity, thrombosis)
- Retinoids: Inhibition of IFN gamma

New treatments:
- Lenalinomide: Better safety profile than thalidomide
- Iberodomide: Clinical trials show some promising result
- Ustekinumab: Promising results (60% improvement in CLASI)
• JAK-STAT inhibitors (block IFN pathway):
  • Ruxolitinib (case series)
  • Baricitinib (14% reduction of activity, similar to Belimumab)

• Blocking IFN I alfa:
  • Sifalimumab: Less flares of disease
  • Anifrolumab (blocks receptor): Clinical trial failed to show efficacy

• Some studies show that IFN kappa is implicated in skin and is a sensor for other IFN and upregulates them

• New strategies would block IFN kappa

• Other strategies:
  • Block pDC cells with molecules (BDCA2)
  • Block TYK2 (used in psoriasis)
In 1991 Dermatomyositis was subdivided according to AutoAbs subtypes: Mi2, SRP, anti-synthetase. Most patients were Negative

Since 2005 new auto-antibodies have appeared and define subsets. These autoantibodies are mutually exclusive

Detection is made through ELISA, Immunoblot, or IP

They are considered markers of DM Syndromes
• Anti-TIF1:
  • Adults: More systemic features, more prominent cutaneous manifestations, Ovoid palatal patch, > 50% malignancies
  • Present in 15-20% of Juvenile DM (most frequent)- No malignancy but ulceration, lipoatrophy, contractures

• Anti-Mi2: Classical DM, good prognosis (less internal involvement, less association with cancer), and good response to tx.

• Anti-NPX2:
  • Juvenile DM: More severe manifestations muscular atrophy, calcinosis, contractures
  • Adults: Calcinosis? Paraneoplastic?

• Anti-MDA5: Mostly associated with amyopathic DM
  • Adult>juvenile
  • More frequent in Asians (Japan- 38% juvenile DM)
Anti-MDA5: Mostly associated with amyopathic DM
- Adult > juvenile
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- High incidence of Interstitial Lung Disease
- Distinct cutaneous phenotype: Ulcerations, palmar papules
- Arthritis
- Little myositis
BP is the most frequent autoimmune subepidermal bullous disease
There is a wide spectrum of clinical presentations: 20% have Atypical BP
These include Non-bullous BP, Pemphigoid nodularis, Lichen planus pemphigoids, Childhood BP, Dyshidrosiform BP, Drug-induced BP, ...
Recently cases of BP induced by Dipeptidyl peptidase-4 inhibitors (gliptins) anti-diabetic drugs and anti-PD1/PD1L Abs are being described
Think of BP in any chronic pruritic eruption \( \Rightarrow \) DIF
However there are not accepted & validated Diagnostic Criteria for BP
When treating the patients always keep in mind that these are elderly and fragile patients with multiple co-morbidities
Avoid the use of high doses of systemic corticosteroids or aggressive immunosuppressive treatments, especially if they have not been validated by rigorous studies