

ACR REVIEW 05 2020 09 NOV

Enfermedades Autoinmunes Sistémicas

Dra. M^a José Cuadrado Lozano
Clínica Universitaria Navarra
Madrid

Earlier Cancer Diagnosis After Idiopathic Inflammatory Myopathy Onset Is Associated with Improved Long Term Survival - Results from Four European Cohorts

- This study aimed to investigate if a shorter time between IIM onset and cancer diagnosis is associated with improved long term survival.
- A total of 248 (68% female) were included in the study
- Cox-proportional hazard modelling indicated that a longer IIM onset to cancer diagnosis time was significantly associated with death for the whole cohort (HR 1.04 [95% CI 1.01, 1.06], p-value < 0.01)
- This relationship was observed in the female cohort only (HR 1.05 [95% CI 1.01, 1.08], p-value < 0.01) and not the male cohort (HR 1.05 [95% CI 0.97, 1.13], p-value 0.26).

Immunostimulatory Herbal Supplement Use Is More Common Among Patients with Dermatomyositis

- The use of complementary and alternative medicine (CAM) is prevalent in dermatology. Certain CAMs, including Spirulina, Alfalfa, Chlorella, Echinacea, and Blue-Green Algae have been reported to incite an immune response or induce dermatomyositis (DM), cutaneous lupus erythematosus (CLE) or autoimmune blistering diseases (AIBD).
- 372 patients were enrolled, including 158 DM (42.5%), 122 CLE (32.8%), 31 AIBD (8.3%), and 61 controls (16.4%).
- CAM use, in particular spirulina, is greater among patients with DM. This association is statistically significant for DM compared to CLE or AIBD.
- Patients with DM should be educated regarding the risk of onset or flare from using immunostimulatory CAM such as Spirulina.

Clinical Characteristics and Outcomes in Patients with Primary Sjögren's Syndrome-Associated ILD

- Features interstitial lung disease (ILD) in up to 16% of patients. The clinical characteristics and natural history of pSS-associated ILD are not well defined.
- 81 patients with pSS-ILD. ILD was the presenting manifestation of pSS in 17 (21%) patients. ILD and pSS were diagnosed concurrently in 34 (43%) patients. In a 36%, the diagnosis was established after a median of 6 years (range 1-31 years)
- New immunomodulatory therapy in 54 (67%) patients.
- Mycophenolate (49%), hydroxychloroquine (44%), azathioprine (25%), and rituximab (7%).
- 21 patients (26%) with pSS-ILD died during follow-up. The median survival time after ILD diagnosis was 11 years (6-12 years).

Sjogren's Syndrome and Its Risk of Cervical Lesions

- The risk of Sjogren's syndrome and cervical malignancy has not been determined due to scarce data.
- Out of the 136 women, 34 (25%) women had abnormal cervical cells
- 25% having concerning pap smears, with 62.3% of these samples infected with high risk HPV strains.
- Sjogren's leads to increased cervical inflammation, as well as susceptibility to HPV-infections that may lead to cervical dysplasia.
- We should offer our Sjogren's patients appropriate HPV vaccination and ensure appropriate cervical cancer screening has been done.

A Panel of Urinary Proteins Predicts Active Lupus Nephritis and Response to Rituximab Treatment

- Presence and/or severity of LN are currently assessed by renal biopsy, but biomarkers in serum or urine samples may provide an avenue for non-invasive routine testing.
- 197 SLE patients and 48 healthy controls were recruited, and urine samples collected. 75 of the SLE patients had active LN and 104 had no or inactive renal disease.
- A combination of five urine proteins, namely LPGDS, transferrin, ceruloplasmin, MCP-1 and sVCAM-1 was a good predictor of active LN (AUC 0.898).
- A combined model of LPGDS, transferrin, AGP-1, ceruloplasmin, MCP-1 and sVCAM-1 predicted response to rituximab treatment at 12 months (AUC 0.818).

Time to Renal Insufficiency Based on 25(OH)-Vit-D Levels

- Vitamin D has multiple immunomodulatory effects. In SLE patients who are vitamin D insufficient/deficient, vitamin D supplementation clearly improves proteinuria.
- There was a significantly elevated risk of renal insufficiency among those in the extremely low levels of 25(OH)-vitamin D (< 20 ng/mL).
- RI was more common in males, African-Americans and obese patients. Results were adjusted for race, age, sex and BMI.
- Data show that very low 25(OH)-vitamin D levels < 20 ng/mL are associated with renal insufficiency

Systemic Treatment in Sarcoidosis. Study of 377 Patients from a Single University Hospital

- 377 patients (188 men/189 women)
- After a mean follow-up of 13.0 ± 9.3 years, 161 (42.7%) patients did not require treatment.
- The remaining 216 received oral glucocorticoids (54.6%) with a maximum mean dose of 43.2 ± 19.0 mg/day, conventional immunosuppressants (16.2%), biological therapy (7.4%) and/or endovenous methylprednisolone (4.0%).
- Biological therapy was indicated by pulmonary (32.1%), ocular (32.1%), neurological (10.7%), muskuloeskeletal (10.7%), cutaneous (7.1%), nephrological involvement (3.6%); and Heerfordt's syndrome (3.6%).
- Adalimumab and Infliximab were the biologics used more frequently