Sjogren’s Syndrome Summary  Dr. Eugene Kissin

Dry eye signs: conjunctival injection, lacrimal enlargement, image irregularity
Tests: Eyes → Schirmer’s, Fluorescein tear break-up, Rose Bengal score. Mouth → unstimulated salivary flow

2016 ACR/EULAR classification criteria (96% sensitive, 95% specific, but does not account for extra-glandular disease)

Exclusion Criteria: Head/neck radiation history, Active Hep C, graft vs. host disease and Sjogren’s mimics which include HIV/AIDS, Sarcoidosis, Amyloidosis, IgG4-related disease

SSA: Found in 1% of healthy women, found in SLE and autoimmune myopathy
SSB without SSA does NOT support Sjogren’s diagnosis
Autoantibodies are present up to 18-20 years before diagnosis of primary Sjogren’s
Lab abnormality prevalence in Sjogren’s patients: ANA (85%), RF (60%), SSA (55%), low C4 (15%), low C3 (1%), Hypergammaglobulinemia (45%), high amylase (25%), anemia (15%), leukopenia (10%), cryoglobulinemia (7%)

Salivary ultrasound can be helpful in increasing diagnostic accuracy → multiple hypoechoic areas and hyperechogenic bands are seen, 83% sensitivity, 91% specificity

Joint symptoms precede sicca in 20% of Sjogren’s patients: 75% respond to HCQ, 92% respond to MTX
Oral lesions: traumatic lesions, oral candidiasis (most common), denture stomatitis, angular cheilitis, median rhomboid glossitis, fissured tongue, aphthae, coated tongue. Pts can develop resistant oral candidiasis, so culture may be necessary for proper treatment.
ILD can be seen in up to 20% of patients and 10-51% will develop ILD before sicca symptoms. Includes NSIP 40%, UIP 10%, COP 4%, LIP 4%, combination in the rest.
Skin changes: flat purpura related to hypergammaglobulinemia, palpable purpura due to vasculitis (higher risk of vasculitis in +RF, SSA, and cryo), urticaria, subcutaneous nodules, skin ulcers
58% with skin disease will develop CNS and peripheral nervous system involvement
Renal disease – tubulointerstitial nephritis 77%, glomerular disease 23% (cryoglobulinemia 9%)
Distal RTA is also commonly seen. Dx with low urinary citrate (risk factor for urolithiasis)
Pulmonary hypertension was noted in 12.5% of Sjogren’s patients and can be first manifestation (Risk factors: pericardial effusion, high titer RF, Raynaud’s liver injury)

Poor prognosis in low C4, cryoglobulins, purpura (best predictor of GN), elevated RF
Lymphoma predictors: parotid enlargement, lymphadenopathy, palpable purpura, low C4, cryoglobulins

Q&A Pearls
-No yearly recommendation of ILD screening in Sjogren’s patients.
-Initially screen Sjogren’s patients with complements and cryoglobulins to help predict extra-glandular disease and closer follow up.
-Check urinary citrate if suspecting RTA initially and if low refer to nephrology for further workup.
-Some data on Omega-3 fatty acids helping with dryness, but no consensus or guidelines.
-If suspicious for sarcoidosis or amyloidosis, biopsy will be required. Ultrasound may show specific findings of diabetes, IgG4, and HIV and biopsy may not be required.
-Low threshold to evaluate unilateral parotid gland swelling with imaging to rule out MALT lymphoma.
-Higher incidence of small fiber neuropathy in Sjogren’s patients compared to healthy controls. After normal EMG/NCS, diagnose with skin biopsy stained for nerves. Begin treatment with Gabapentin, Elavil, and consider IVIG for worse cases.