Lessons from Sjogren’s Syndrome
Dry eyes and mouth in Scleroderma

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What is Sjogren’s syndrome?

Chronic autoimmune disease characterised by inflammation in saliva and tear glands

Cause unknown

First described in literature 1892 (Mikulicz) & 1925 (Gougerot)

Henrik Sjogren published his doctoral thesis 1933
Prevalence

Primary SS - 0.1 – 0.4%

Secondary SS - 10-20% of patients with longstanding RA

Overlaps with other conditions eg SLE & scleroderma

Presentation often delayed & non-specific

Often ‘grittiness’ rather than dry eye

Oral symptoms may require prompting
Making the Diagnosis!

Ocular symptoms
Oral symptoms
Ocular signs
Oral signs
Histopathology
Anti-Ro/La antibodies

American-European consensus group (AECG) criteria
Ann Rheum Dis, 2002; 61: 554-8
Glandular features

Changes in amount & composition of secretions resulting in:

Dry eyes
Dry mouth
Dry skin
Dry vagina
Dry cough
General advice

Avoid dry atmospheres
Humidify rooms
Chew sugarless chewing gum
Shower rather than bath & avoid soap
Wear glasses (with side arms)
Avoid too much PC use (reduces blink rate) &
laser surgery!
Dry eyes

Burning, grittiness
Fatigue, blurring
Watery eyes – due to increased reflex tearing in early stages
Hypoanaesthetic eye surface in later disease (avoid laser surgery)
Marginal tear film (norm ~2mm) reduced
Break up time reduced (norm >10secs)
Tear Film

Lipid layer
0.1 μm

Lacrimal gland

Aqueous layer
7.0 μm

Meibomian glands

Goblet cells

Mucous layer
0.2 μm

Values

Service Teamwork Ambition Respect
Topical treatment of dry eye

Start with simple lubricants

Hypromellose 0.3%

Carbomer gel e.g. Clinitas 0.2% gel

Carmellose e.g. Optive 0.5% (Biodegradable preservative)

Switch to preservative free if eye drop use > 4x daily
Topical treatment of dry eye

Use longer acting agents next
Carbomer e.g. clinitas gel 0.2%
Carmellose e.g. celluvisc 0.5% SDU
Sodium hylauronate e.g. Hylo-Tear 0.1% or Hylo-Forte 0.2%
VitA-Pos eye ointment at night
If mucus stranding use mucolytics e.g. acetylcysteine eye drops
Treatment of dry eye

Consider punctal occlusion (temporary first)

Stimulate meibomian gland secretions & use liposomal sprays

Treat blepharitis (warm compresses, bicarbonate, topical antibiotics, oral doxycycline, omega 3 supplements)
Treatment of dry eye

Autologous serum ‘tears’
Cyclosporin eye drops and ointment
Steroid eye drops
Dry mouth

Increased caries & tooth decay
Oral candida (white or red)
Angular cheilitis
Bad breath
Dysphagia
Topical treatment of dry mouth

Water

Avoid sugared, acidic or fizzy drinks

Chew sugarless chewing gum

Biotene Oralbalance salivary gel, BioXtra, Salivaorthana & Luborant all contain fluoride

Glandosane fluoride free & acidic – so avoid

Lozenges/pastilles no good if very dry
Topical treatment of dry mouth

Toothpastes (eg Duraphat) and mouthwashes

Home made mouthwash (1 tsp salt & 1 tsp baking powder in 1 litre water)

Olive oil, yoghurt

Treat candida if present (fluconazole 50mg orally or nystatin liquid or amphotericin lozenges for max 10/7 then break)
Vaginal dryness

Simple lubricants e.g. KY jelly

Non-hormonal moisturisers e.g. Replens, Sylk, Repadina (Hyaluron based capsules), vagisan

Oestrogen creams eg vagifem

HRT
Pilocarpine (salagen)

Muscarinic acetylcholine receptor agonist

May be more beneficial early rather than late

Start with low dose e.g. 5mg per day or consider drops (4% pilocarpine eye drops, 1 drop ~ 2mg)

Increase dose slowly to facilitate development of tolerance to side effects

Persevere with treatment to see effects
Fatigue

Physical & mental fatigue affects at least 75% of patients with Sjogren’s

Significantly higher levels of fatigue than healthy controls

Similar levels of mental & physical fatigue to patients with SLE

Bowman et al 2004
Theander et al 2010
Fatigue

Fatigue most disabling symptom for many
May correlate with sleep disturbance & anxiety
Influenced by pre-morbid personality
Encourage exercise
Improve sleep quality
Don’t forget other causes (eg anaemia, hypothyroid, depression, stress)
Mental Health

Sjogren’s patients have high levels of ‘psychological distress’ compared to Healthy controls.

Personality features associated with SjS include negativity, preoccupation with detail, perfectionism & anxiety.

Karaiskos et al Rheumatology 2010
Skin

Skin dryness affects >50%

Photosensitive rashes common in Ro+

Sub- acute cutaneous lupus in Ro+ve

Ensure UVA and UVB protection (look for high protection factors and star rating)

Davidson et al 1999
Urinary symptoms

Mild symptoms of urinary dysfunction in 61% of patients with SS (cf 27% controls)

Severe symptoms 14% SS (cf 7% controls)

27% of SS patients report urinary frequency & 36% complain of suprapubic pain

*Haarala et al 2000*
Urinary Tract Infection

Probable increased frequency of UTI

Freq of UTI 6% in RA alone, 30% in RA/SS

Tischler et al 1992
Interstitial Cystitis

Chronic inflammatory bladder disease
90% females
Chronic inflammation of bladder wall
No evidence of infection
Cause unknown
Associated with underlying CTD
23% of patients with IC may have Sjögren’s

Van der Merwe et al 2003
Irritable Bowel Syndrome

Bloating, diarrhoea, abdominal colic

Affects 20% of normal population

Prevalence in Sjogren’s unknown but anecdotally common

‘Slow motility’ often observed
Management of Systemic Disease

Fatigue often benefits from graded exercise programme

Mindfullness

Hydroxychloroquine
Take Home Message

Sjögrens syndrome may overlap with Scleroderma
Treat dryness early
Meticulous dental hygiene
Make friends with your dentist!
Further advice/help www.bssu.uk.net