

# Sindromul Sjogren

Dr.Camelia Badea

Spitalul clinic Colentina

Medicina Interna 2

# Sindrom Sjogren

- afectiune inflamatorie cronica
- afecteaza glande exocrine
- manifestari extraglandulare

\* Manifestari clinice frecvente – xerostomie, xerooftalmie

# Epidemiologie

- Incidenta: 7/100000
- Prevalenta :43/100000
- Predominanta: feminina – raport 9/1
- Debut frecvent – decada a-5a

# Etiopatogenie

1. Sindrom Sjogren primar
2. Sindrom Sjogren secundar – asociat unor alte boli de colagen
  - Poliartrita reumatoida 7-25%
  - LES – 9 – 14%
  - BMTC
  - Dermatopolimiozita
  - Sclerodermie
  - Crioglobulinemie

# Etiopatogenie

1. Factori genetici – agregari familiale cu bol autoimmune
2. Factori de mediu: - infectii EBV, CMV, coxsackie, HTLV 1, retrovirusuri etc

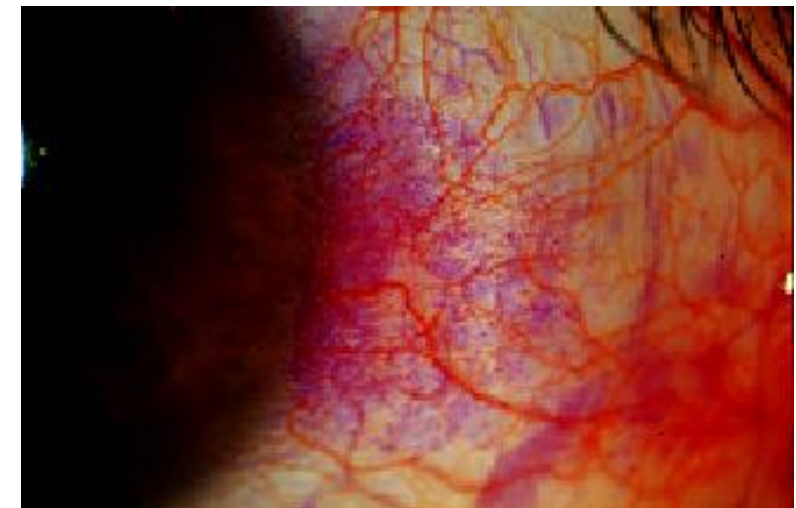
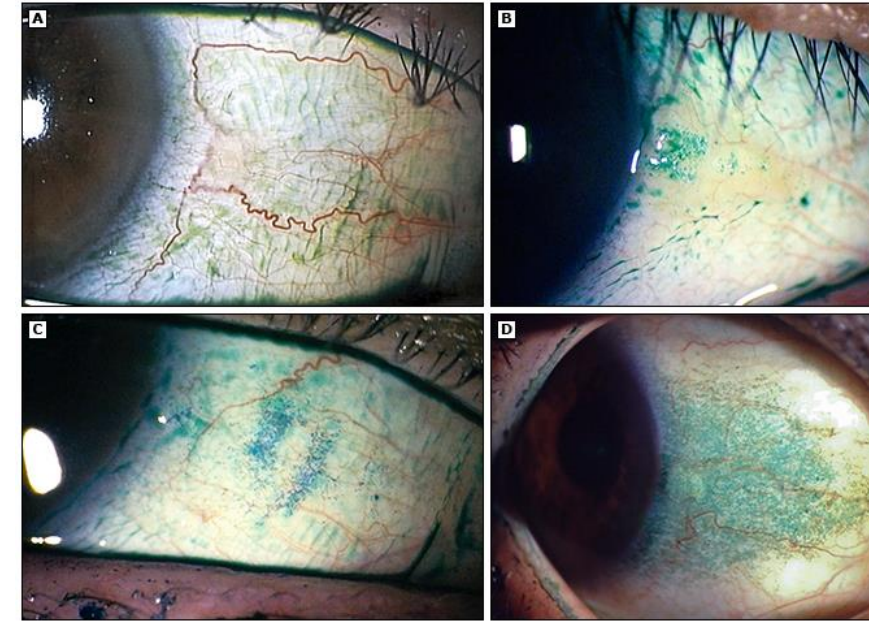
- infectiile virale sunt considerate declansatoare a reactiei immune la nivel glandular prin mecanism de “mimetism molecular”

- infiltrare limfocitara predominant LT CD4+ la nivelul glandelor

# Manifestari clinice

## 1. Afectare oculara:

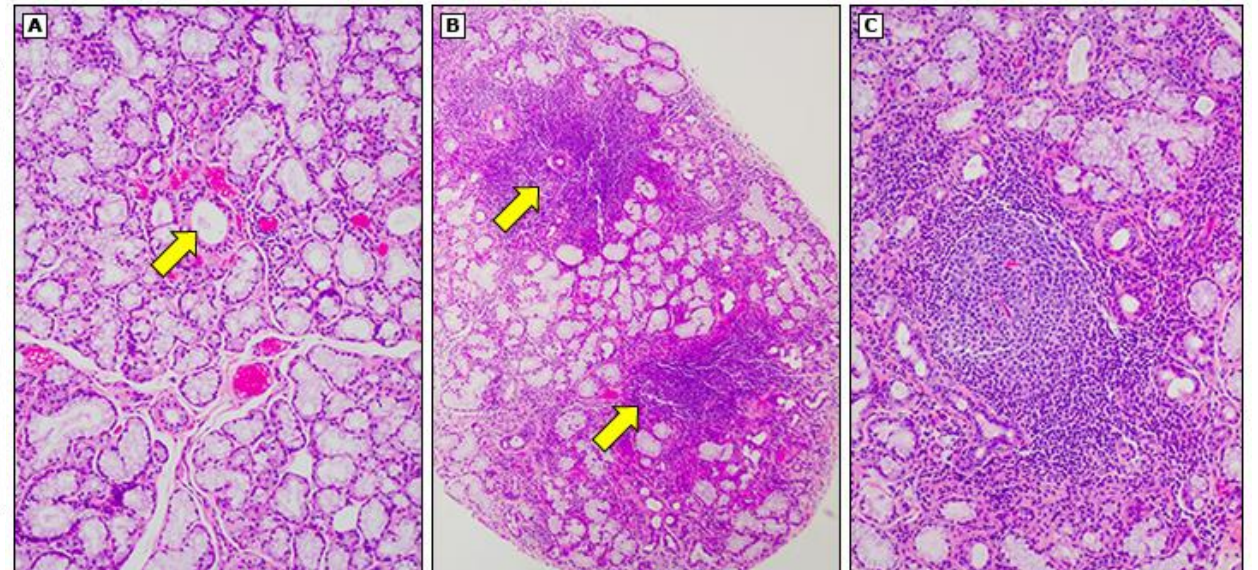
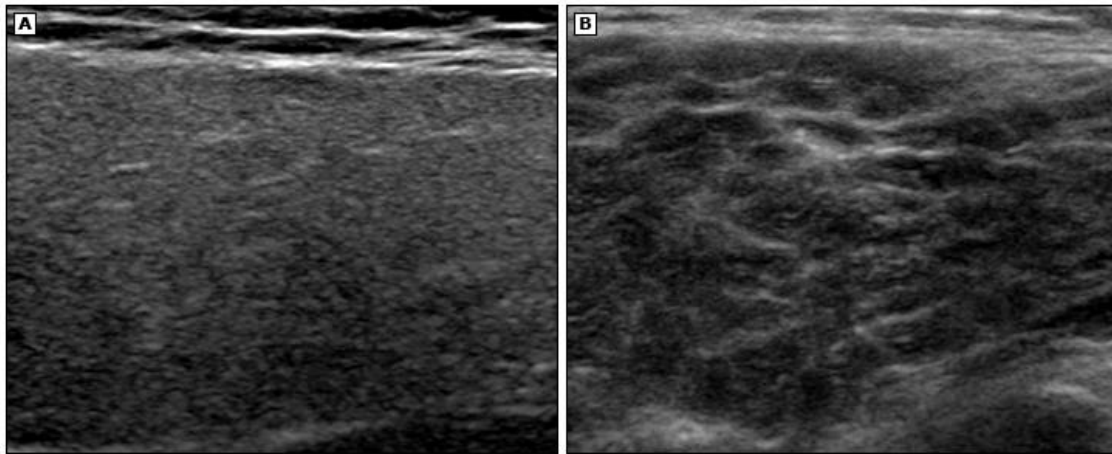
- keratocojunctivita sicca – uscaciune, arsura, senzatie de “nisip in ochi”, fotofobie
- clinic: conjunctive congestionate, eroziuni corneene, suprainfectii, marire in volum a glandelor lacrimale
- Test Schirmer: <5mm – pozitiv
- test roz bengal
- de exclus consumul de medicamente anticolinergice – pot da test fals pozitiv



## 2. Afectare salivara:

- xerostomie, disfagie, dificultate de a vorbi
- candidoza orala, infectii dentare – edentatie
- pierderea senzatiei gustative prin atrofia papilelor
- 30-60% - tumefierea glandelor parotide
- pot fi sediul transformarii in limfom

- Biopsia de glanda salivara minora – “standard de aur” – identifica infiltrate limfoplasmocitar periacinar
- Biopsie pozitiva cand scor focus  $>1$
- Focus = o aglomerare de minim 50 limfocite,
- Scorul focus = nr de focus-uri pe o suprafata de 4 mm
- Alte teste: sialografie,scintigrama, ecografie





- Alte afectari glandulare:

- pulmonar: uscaciunea mucoasei nazale, scaderea secretiilor bronsice
- digestiv: disfagie, hipoclorhidrie, hipopepsinogenemie (gastrita atrofica)
- pancreas: - rara pancreatita autoimuna
- vagin: uscaciune

# Afectare extraglandulara

1. Semne generale: astenie, febra, scadere in greutate
2. Afectare musculoscheletala: artralгии, artrite, mialгии difuze
3. Afectare cutanata: xeroza, prurit, eruptii eritemato-papuloase, eritem anular
4. Manifestari vasculitice cutanate: vasculita leucocitoclazica – purpura palpabila, rar leziuni ischemice
5. Sindrom Raynaud

6. Afectare pulmonara: leziuni fibrotice interstitiale – pneumonie interstitiala limfocitara, BOOP
7. Afectare renala – nefrita interstitiala si acidoza tubulara
8. Afectare neurologica: neuropatie periferica senzitiva, mixta, mononeuritis multiplex, neuropatie vegetative
9. Risc de transformare maligna – LMNH - >44x (predispozitia – adenopatie, tumefiere parotidiana persistenta, purpura, hipocomplementemie, crioglobulinemie)

# Paraclinic

## Hematologic:

- leucopenie usoara cu limfopenie
- trombocitopenie moderata
- anemie normocroma normocitara

## Imunologic:

- hipergamaglobulinemie + crioglobulinemie
- FR pozitiv
- ac anti Ro (SSA)
- ac anti La (SSB)

# Criterii Consensului Americano-European

1. Simptome de ochi uscat (>3luni)
2. Semne ochi uscat: T Schirmer, roz Bengal
3. Simptome de gura uscata (>3 luni)
4. Semne de gura uscata: sialografie, scialometrie, scintigrama
5. Biopsie gl salivara minora – pozitiv
6. Prezenta ac anti Ro, La

Diasgnostic 4/6 (obligatoriu unul din 5,6)

## Criteria de excludere:

- radioterapie la nivel cap, gat
- infectie VHC
- infectie HIV/SIDA
- sarcoidoza
- antecedente de limfom
- medicatie anticolinergica

# Diagnostic diferential

1. Scaderea secretiilor - consum de betablocante, clonidine, antispastice (papaverina), antihistaminice, antidepresive etc
2. Xerostomie – folosirea lentilelor de contact
3. Amiloidoza
4. Infectii VHC,HIV, etc

# Tratament

1. Masuri igenodietetice – igiena riguroasa, evitarea alimente astringent
2. Tratament simptomatic
  - xerostomie: agonisti colinergico – pilocarpine, cevimeline (efect stimulat pe receptorii muscarinici glandulari)
  - xerooftalmie: lacrimi artificiale, local – picaturi cu corticosteroizi, ciclosporina



# Tratament

3. Afectare articulara: AINS, doze mici de cortizon, hidroxicloroquine

4. Afectare extraglandulara:

- medicatie imunosupresoare: ciclofosfamida, Imuran, micofenolat mofetil ( vasculita, afectare pulmonara, neuropatii)

- medicatie biologica – rezultate neconcludente:

- Rituximab ( ac monoclonal anti CD 20)

- belimumab (ac monoclonali anti BAFF)

- abatacept