Nonneoplastic Diseases of the Salivary Glands

Frederick S. Rosen, MD
Faculty Advisor: Francis B. Quinn, MD
The University of Texas Medical Branch
Department of Otolaryngology
Grand Rounds Presentation
October 3, 2001
Introduction

- Two major categories: inflammatory and noninflammatory
- Usually involve major salivary glands
- Usually do not require extensive diagnostic workup
Inflammatory Diseases

- Mumps
  - Most common viral disorder of salivary glands
  - Peak age 4-6
  - 1 or both parotids after 2-3 week prodrome
  - Diagnosis: serology or urine
  - Complications: deafness, pancreatitis, meningitis, orchitis, Type I DM, chronic obstructive sialadenitis
Inflammatory Diseases

- **Other Viruses**
  - CMV, Coxsackievirus A, Echovirus, Influenza A, Lymphocytic choriomeningitis Virus
  - Treatment: symptomatic for all viral diseases

- **Acute Suppurative Sialadenitis**
  - Parotid most common site; peak age 50’s-60’s
  - 30-40% in post-op patients; most commonly GI procedures POD 3-5
Inflammatory Diseases

- Acute Suppurative Sialadenitis
  - Presentation: sudden, diffuse enlargement with associated induration and tenderness. Massage produces purulent saliva
  - 20% of cases bilateral
  - Pathogens: *Staph aureus most common*; Gram negatives, anaerobes also common
  - Treatment: hydration, improved oral hygiene, repeated massage of gland, IV abx, warm compresses, sialogogues
Inflammatory Diseases

Acute Suppurative Sialadenitis
- If no significant improvement in 24-48h, then proceed to incision & drainage OR image-guided needle aspiration

Chronic Sialadenitis
- Most commonly parotid
- Usually from permanent damage during acute infection; occasionally from recurrent parotitis of childhood
Inflammatory Diseases

- **Chronic Sialadenitis**
  - Histologic changes: sialectasis, progressive acinar destruction, lymphocytic infiltrates
  - Saliva changes; returns to normal between attacks
  - Presentation: mild pain, recurrent parotid enlargement that worsens with eating; 80% develop xerostomia
Inflammatory Diseases
Inflammatory Diseases

- Chronic Sialadenitis
  - Treatment
    1) Underlying causes
    2) Sialogogues, massage, heat, hydration, abx during acute attacks
    3) Periodic ductal dilatation, duct ligation, total gland irradiation, tympanic neurectomy
    4) Excision
Inflammatory Diseases

- Recurrent Parotitis of Childhood
  - More common in males; peak age 5-7
  - ¾ give h/o Mumps; heredity plays no role
  - Presentation: Usually unilateral; when bilateral, one side worse
  - Severe pain, fever, malaise during attacks
  - Recurs
Inflammatory Diseases

- Recurrent Parotitis of Childhood
  - Disease course (Ericson): onset age 3 months - 16 years
  - Exacerbations every 3-4 months
  - 55% of cases resolve with puberty
  - 25% no improvement with puberty
  - Histology: massive B-cell infiltration and dilated intraglandular ducts
Inflammatory Diseases
Inflammatory Diseases

- Recurrent Parotitis of Childhood
  - Sialogram: multiple peripheral sialectases 1-2 mm in diameter; changes persist w/ resolution of symptoms
  - Pathogens: flora ascend from oral cavity
    - Balls of soft material common, but rarely yields frank pus
  - Treatment: Pen VK, massage, warmth, good oral hygiene, sialogogues, chewing gum
Inflammatory Diseases
Inflammatory Diseases

- Benign Lymphoepithelial Lesion
  - Epimyoepithelial islands arise from
    1) lymphoreticular infiltrates
    2) acinar atrophy
    3) ductal metaplasia
  - Presentation: Asymptomatic enlargement of 1 gland
  - Risk of lymphoma, carcinoma, pseudolymphoma
  - No treatment necessary
Primary Tuberculosis

- Presentation: Unilateral parotid
  May present as acute inflammatory lesion or as chronic tumorous lesion
- Diagnosis: AFB stain of saliva AND PPD test
  FNA if tumorous lesion
- Treatment: Anti-TB meds; excision if resistant
- Secondary TB: systemic dz.; submandibular and sublingual glands more often involved
Inflammatory Diseases
Inflammatory Diseases

- Animal Scratch Disease
  - Typically attacks periparotid lymph nodes
  - Pathogens: *Bartonella henselae, Afipia felis*
  - Diagnostic Criteria (3/4):
    1) H/o contact w/ a cat and presence of scratch
    2) + skin test or + serology for *B henselae*
    3) + Gram stain and Cx
    4) Histology: stellate abscesses, pleomorphic intracellular bacilli, Warthin-Starry stain
Inflammatory Diseases

- Animal Scratch Disease
  - Should place PPD to r/o Tb
  - 96% resolve spontaneously within 2-6 months; close followup needed until adenopathy subsides
  - Treatment: Bactrim X 1 week, or Rifampin X 1-2 weeks
  - IV Gentamicin in severe cases
**Inflammatory Diseases**

- **Actinomycosis**
  - Infection from tonsil or teeth
  - Presentation: 61% visible sinus tracts; 40% adenopathy; some have purplish skin discoloration
  - Histology: sulfur granules
  - Diagnosis: culture
  - Treatment: I&D, 2-6 weeks of IV Pen G
Inflammatory Diseases
Inflammatory Diseases

Atypical (Nontuberculous) Mycobacteria
- Median age 28 months (usually ages 1-5)
- Transmitted from soil to mouth/eyes
- Presentation: focal swelling of face or neck (100%), change in overlying skin color (76%), necrosis or fistula of skin (52%); no systemic symptoms
- Most common sites: submandibular area, parotid, upper neck, submental area
Inflammatory Diseases

- Atypical (Nontuberculous) Mycobacteria
  - Pathogen: *MAC* most common
  - Diagnosis: FNA diagnostic 87% of time; PPD’s not helpful
    1) Culture from FNA, or
    2) + AFB stain from FNA, or
    3) Histology: granulomatous inflammation w/ caseating necrosis
Inflammatory Diseases
Inflammatory Diseases

- Atypical (Nontuberculous) Mycobacteria
  - Treatment: Curettage vs. Excision
    - Curettage for lesions with extensive skin necrosis or fluctuant parotid lesions
    - Surgical excision more effective
  - Medications controversial; Macrolides may work for early disease
Inflammatory Diseases

- Sarcoidosis
  - 6% involve salivary glands clinically, 1/3 histologically
  - Heerfordt’s syndrome (Uveoparotid fever):
    1) Uveitis
    2) Parotid enlargement
    3) CN VII paralysis

Self-limited; uveitis can result in glaucoma – requires long term f/u
Inflammatory Diseases

Sjogren’s Syndrome: Background

- Chronic, slowly progressive, benign; 2\textsuperscript{nd} most common autoimmune disease behind RA
- Lymphocyte-mediated destruction of exocrine glands producing keratoconjunctivitis sicca and xerostomia
- 90\% middle-aged women
- 44\% report PCN allergy
Inflammatory Diseases

Sjogren’s Syndrome: Background

- Primary=exocrine glands only; Secondary=coexisting autoimmune disease
- Secondary form more common; salivary gland enlargement more common in primary form
- Serology (similar pattern in SLE):
  1) ANA (50-80%)
  2) RF (75%)
  3) Ro/SS-A antibodies
  4) La/SS-B antibodies; 3 or 4 in up to 90%
Inflammatory Diseases

- Sjogren’s Syndrome: Presentation
  - Xerostomia: most bothersome; difficulty swallowing dry food, difficulty speaking continuously, burning sensation, increased caries, problems wearing dentures; erythematous/sticky oral mucosa, atrophy of filiform papillae
  - Keratoconjunctivitis Sicca: gritty feeling under eyelids, blurred vision, burning sensation, thick strands at inner canthi, decreased tearing, redness/itching, photosensitivity; results from destruction of conjunctival epithelium
Inflammatory Diseases

Sjogren’s Syndrome: Presentation
- Other exocrine gland involvement: dry nose, dry throat, xerotrachea, esophageal mucosal atrophy, atrophic gastritis, subclinical pancreatitis, vaginal dryness
- 1/3 = fatigue, low grade fever, myalgias/arthralgias
- Extraglandular involvement in ¼: Lungs, kidneys, vasculitis, nervous system
Inflammatory Diseases

Fig. 15–3. Other glandular manifestations observed in Sjogren’s syndrome patients from a multicenter study for the diagnostic criteria for Sjogren’s syndrome, Pisa, 1992. (626 cases; 19 European centers)
Inflammatory Diseases

- Sjogren’s Syndrome: Associated risks
  - Increased risk of
    1) NonHodgkin’s Lymphoma (RR=44)
    2) Multiple Myeloma
Inflammatory Diseases

Table 15-4. Preliminary criteria for the classification of Sjogren's syndrome (modified from Reference 9).

1. Ocular symptoms
   A positive response to at least 1 of the following 3 questions:
   (a) Have you had daily, persistent, troublesome dry eyes for more than 3 months?
   (b) Do you have a recurrent of sandy or gravel feeling in the eyes?
   (c) Do you use tear substitutes more than 3 times a day?

2. Oral symptoms
   A positive response to at least 1 of the following questions:
   (a) Have you had a daily feeling of dry mouth for more than 3 months?
   (b) Have you had recurrent or persistently swollen salivary glands as an adult?
   (c) Do you frequently drink liquids to aid in swallowing dry foods?

3. Ocular signs
   Objective evidence of ocular involvement determined on the basis of a positive result on at least 1 of the following 2 tests:
   (a) Schirmer-1 test (≤ 5 mm in 5 minutes)
   (b) Rose bengal score (≥ 4, according to the van Bijsterveld scoring system)

4. Salivary gland involvement
   Objective evidence of salivary gland involvement, determined on the basis of a positive result on at least 1 of the following 3 tests:
   (a) Salivary scintigraphy
   (b) Parotid sialography
   (c) Unstimulated salivary flow (≥ 1.5 ml in 15 minutes)

5. Histopathologic findings
   Focus score ≥ 1 on minor salivary gland biopsy
   (focus defined as an agglomeration of at least 50 mononuclear cells, focus score defined as the number of foci/4mm² of glandular tissue)

6. Autoantibodies
   Presence of at least 1 of the following autoantibodies in the serum:
   Antibodies to Ro (SS-A) or La (SS-B) antigens or antinuclear antibodies or rheumatoid factor.

A patient is considered as having probable Sjogren’s syndrome if 3 of 6 criteria are present, and as definite if 4 of 6 criteria are present.
Inflammatory Diseases

- **Sjogren’s Syndrome: Histology**
  - Severe lymphoid (T-cell) infiltrate can mimic lymphoma; heterogenous, lobular architecture preserved
  - Enlarged lymph nodes w/ pleomorphic infiltrates and frequent mitotic figures = “pseudolymphoma”
  - When biopsying, avoid epinephrine; send specimen in formalin
Inflammatory Diseases
Inflammatory Diseases

- Sjogren’s Syndrome: Treatment
  - Incurable disease
  - Key=fluid replacement
    - Artificial tears; eye patching, boric acid ointments for corneal ulceration
    - Avoid diuretics, antihypertensives, antidepressants
  - Medications: Pilocarpine 5 mg TID; hydroxychloroquine; glucocorticoids 1 mg/kg/day
Noninflammatory Diseases

Sialolithiasis

- 80% submandibular gland, 20% parotid
- Only 1 stone in ¾ cases
- 90% of submandibular stones radioopaque; 90% of parotid stones radiolucent
- Presentation: recurrent swelling, pain worse with eating
- Complications: sialadenitis, ductal ectasia, and stricture
- Treatment: If near duct orifice, transoral removal of stone with marsupialization
  If near hilum, gland excision
Inflammatory Diseases
Noninflammatory Diseases

- Cysts
  - Mucoceles vs. Mucous cysts: minor salivary glands
  - 2-5% of all parotid lesions
  - Congenital: dermoid cysts, ductal cysts, 1st arch branchial cleft cysts
  - Acquired: BLL, trauma, parotitis, calculi, neoplasms
Noninflammatory Diseases

Trauma

- Identify the duct; can pass probe to ID distal duct; can milk gland to ID proximal duct
- Transected duct: end-to-end anastomosis over polyurethane catheter with 9-0 suture; remove catheter after 2 weeks
- Salivary-cutaneous fistula: repeat aspiration and pressure dressings; sialogram; excision if conservative treatment fails
- Blunt trauma: drain large hematomas early
Noninflammatory Diseases

- **Sialadenosis**
  - Nonneoplastic, noninflammatory enlargement of salivary glands associated with systemic disorders
  - Usually asymptomatic
  - Causes=obesity, malnutrition, malabsorption, and alcoholic cirrhosis; very rarely does sialadenosis occur in nonalcoholic cirrhosis
Noninflammatory Diseases

- **Cheilitis Glandularis**
  - Enlargement of the labial salivary glands; clear, thick, sticky mucus; can result in lower lip eversion
  - Treatment: vermilionectomy

- **Kussmaul’s Disease (Dialodochitis Fibrinosa)**
  - Mucous plug obstructing duct
  - Treatment: rehydration, gentle massage, sialogogues
Noninflammatory Diseases

- **Necrotizing Sialometaplasia**
  - Benign, self-healing process of unknown etiology
  - Presentation: usually hard palate, usually males; asymptomatic mucosal ulceration
  - Histology: easily mistaken for SCCA, mucoepidermoid CA; lobular necrosis + squamous metaplasia + preserved lobular architecture
  - Treatment: biopsy for diagnosis, but treatment unnecessary

- Subacute necrotizing sialadenitis = painful, nodular variant