Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

*Hint: NOT ‘benign’ and ‘malignant’—think more broadly than that!*
Lacrima! Gland Enlargement

Enlarged lacrimal gland

- Neoplastic
- Inflammatory

Two broad categories in the differential
Lacrimal Gland Enlargement

Enlarged lacrimal gland

- Neoplastic
  - Two broad categories in the differential

- Inflammatory
  - Two broad categories in the differential

*NOT* 'benign' and 'malignant'—again, think more broadly than that!
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Lymphoid

Inflammatory

Two broad categories in the differential
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Two broad categories in the differential

Lymphoid

Inflammatory

Two broad categories in the differential
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial
  - Two broad categories in the differential
  - Benign (Finally) Malignant

Lymphoid
  - Two broad categories in the differential

Inflammatory
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign

Malignant

Lymphoid

Inflammatory

Most common within each category?
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Malignant

Adenoid cystic carcinoma

Lymphoid

Two broad categories in the differential

Inflammatory

Most common within each category?
What is the classic pathologic description of adenoid cystic carcinoma?
What is the classic pathologic description of adenoid cystic carcinoma?
It looks like ‘Swiss cheese’ owing to its cystic nature.

Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

- Epithelial
  - Benign
    - Pleomorphic adenoma
  - Malignant
    - Adenoid cystic carcinoma

Inflammatory

- Lymphoid
  - Two broad categories in the differential

Two broad categories in the differential

What is the classic pathologic description of adenoid cystic carcinoma?
It looks like ‘Swiss cheese’ owing to its cystic nature.
What is the classic pathologic description of adenoid cystic carcinoma?
It looks like ‘Swiss cheese’ owing to its cystic nature

How is the diagnosis confirmed?
What is the classic pathologic description of adenoid cystic carcinoma?
It looks like ‘Swiss cheese’ owing to its cystic nature

How is the diagnosis confirmed?
Via incisional biopsy
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Malignant

Adenoid cystic carcinoma

Inflammatory

Lymphoid

Two broad categories in the differential

What is the classic pathologic description of adenoid cystic carcinoma?
It looks like ‘Swiss cheese’ owing to its cystic nature

How is the diagnosis confirmed?
Via incisional biopsy

There are five subtypes based on histology. All are bad, but which conveys the worst prognosis?
What is the classic pathologic description of adenoid cystic carcinoma?
It looks like ‘Swiss cheese’ owing to its cystic nature.

How is the diagnosis confirmed?
Via incisional biopsy.

There are five subtypes based on histology. All are bad, but which conveys the worst prognosis? Basaloid.
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Inflammatory

Lymphoid

Two broad categories in the differential

If pleomorphic adenoma is suspected, should incisional biopsy be performed?

No! The tumor should be removed en bloc, without violating its pseudocapsule. Why must a pleomorphic adenoma be managed in this way? Incomplete excision increases the risk of recurrence, which can be malignant (a process known as malignant transformation).
Lacrima; Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Malignant

Inflammatory

Lymphoid

If pleomorphic adenoma is suspected, should incisional biopsy be performed? No! The tumor should be removed en bloc, without violating its pseudocapsule.

Two broad categories in the differential

Two broad categories in the differential

Two broad categories in the differential

Two broad categories in the differential
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Malignant

Neoplastic Inflammatory Epithelial Lymphoid

Benign Pleomorphic adenoma

Adenoid cystic carcinoma

Benign lymphoproliferative disease

Two broad categories in the differential

Inflammatory

Lymphoid

If pleomorphic adenoma is suspected, should incisional biopsy be performed?
No! The tumor should be removed en bloc, without violating its pseudocapsule

Why must a pleomorphic adenoma be managed in this way?
Lacrimal Gland Enlargement

If pleomorphic adenoma is suspected, should incisional biopsy be performed? No! The tumor should be removed en bloc, without violating its pseudocapsule.

Why must a pleomorphic adenoma be managed in this way?
Incomplete excision increases the risk of recurrence, which can be malignant (a process known as malignant transformation).
If pleomorphic adenoma is suspected, should incisional biopsy be performed?
No! The tumor should be removed en bloc, without violating its pseudocapsule.

Why must a pleomorphic adenoma be managed in this way?
Incomplete excision increases the risk of recurrence, which can be malignant (a process known as malignant transformation).
Lacrimal Gland Enlargement

Two broad categories in the differential

Enlarged lacrimal gland

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Malignant

Adenoid cystic carcinoma

NEVER biopsy to make dx

Fungal Thyroid eye dz

Inflammatory

Two broad categories in the differential

Lymphoid

Hold on! How can you know beforehand which you’re dealing with, and therefore whether or not to biopsy?
Lacrimal Gland Enlargement

Two broad categories in the differential

Enlarged lacrimal gland

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Malignant

Adenoid cystic carcinoma

NEVER biopsy to make dx

Inflammatory

Lymphoid

Biopsy to make dx

Hold on! How can you know beforehand which you’re dealing with, and therefore whether or not to biopsy? These entities are usually distinguishable on the basis of Radiographic Features and Clinical Presentation:
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign
- Pleomorphic adenoma
- Adenoid cystic carcinoma
  - NEVER biopsy to make dx

Malignant

- Adenoid cystic carcinoma: 'Bony involvement'
- Pleomorphic adenoma: 'Well circumscribed'
  - Biopsy to make dx

Inflammatory

Lymphoid

Two broad categories in the differential

Hold on! How can you know beforehand which you’re dealing with, and therefore whether or not to biopsy? These entities are usually distinguishable on the basis of Radiographic Features and Clinical Presentation:
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign

Malignant

Pleomorphic adenoma

Adenoid cystic carcinoma

NEVER biopsy to make dx

Biopsy to make dx

Hold on! How can you know beforehand which you’re dealing with, and therefore whether or not to biopsy?

These entities are usually distinguishable on the basis of Radiographic Features and Clinical Presentation:

Pleomorphic adenoma: ‘Well circumscribed’

Adenoid cystic carcinoma: ‘Bony involvement’
Lacrimai Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Benign

Malignant

Pleomorphic adenoma

Adenoid cystic carcinoma

Biopsy to make dx

NEVER biopsy to make dx

INflammatory

Lymphoid

Two broad categories in the differential

Hold on! How can you know beforehand which you’re dealing with, and therefore whether or not to biopsy? These entities are usually distinguishable on the basis of Radiographic Features and Clinical Presentation:

Pleomorphic adenoma: ‘Well circumscribed’

Adenoid cystic carcinoma: ‘Bony involvement’

Pleomorphic adenoma: (rate of growth) and (pain?)

Adenoid cystic carcinoma: (rate of growth) and (pain?)
Hold on! How can you know beforehand which you’re dealing with, and therefore whether or not to biopsy? These entities are usually distinguishable on the basis of Radiographic Features and Clinical Presentation:

- **Pleomorphic adenoma:** ‘Well circumscribed’
- **Adenoid cystic carcinoma:** ‘Bony involvement’
- **Pleomorphic adenoma:** ‘Slow growing’ and ‘Painless’
- **Adenoid cystic carcinoma:** ‘Rapid growth’ and ‘Painful’
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Malignant

Adenoid cystic carcinoma

Lymphoid

Two broad categories in the differential

Inflammatory

Next…
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Benign

Malignant

Pleomorphic adenoma

Adenoid cystic carcinoma

Lymphoid

Benign

Malignant

Two broad categories in the differential

Inflammatory
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial
- Benign
  - Pleomorphic adenoma
- Malignant
  - Adenoid cystic carcinoma

Lymphoid
- Benign
- Malignant

Inflammatory

Two broad categories in the differential

Most common within each category?
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Malignant

Adenoid cystic carcinoma

Lymphoid

Benign

lymphoproliferative disease

Malignant

Lymphoma

Most common within each category?
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign
  Pleomorphic adenoma

Malignant
  Adenoid cystic carcinoma

Lymphoid

Benign
  Benign lymphoproliferative disease

Malignant
  Lymphoma

Inflammatory

Two broad categories in the differential

Next…
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic
- Epithelial
  - Benign
    - Pleomorphic adenoma
  - Malignant
    - Adenoid cystic carcinoma
- Lymphoid
  - Benign lymphoproliferative disease
  - Malignant lymphoma

Inflammatory
- Infectious
- Noninfectious
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign

Malignant

Pleomorphic adenoma

Adenoid cystic carcinoma

Lymphoid

Benign

Malignant

Benign lymphoproliferative disease

Lymphoma

Inflammatory

Infectious

Noninfectious

Two broad categories in the differential

Types of infections, not specific bugs

Benign

Malignant

Lymphoma
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Malignant

Adenoid cystic carcinoma

Lymphoid

Benign

Benign lymphoproliferative disease

Malignant

Lymphoma

Inflammatory

Infectious

Viral

Noninfectious

Bacterial

Fungal
Lacrimal Gland Enlargement

Enlarged lacrimal gland

- Neoplastic
  - Epithelial
    - Benign
      - Pleomorphic adenoma
    - Malignant
      - Adenoid cystic carcinoma
  - Lymphoid
    - Benign
      - Benign lymphoproliferative disease
    - Malignant
      - Lymphoma

- Inflammatory
  - Infectious
    - Viral
      - Which virus is the most common cause of acute dacryoadenitis?
  - Noninfectious

Two broad categories in the differential

- Two broad categories in the differential
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign
- Pleomorphic adenoma
- Adenoid cystic carcinoma

Malignant

Lymphoid

Benign
- Benign lymphoproliferative disease

Malignant
- Lymphoma

Two broad categories in the differential

Inflammatory

Infectious

Viral

Which virus is the most common cause of acute dacryoadenitis?

Epstein-Barr virus
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic
  - Epithelial
    - Benign
      - Pleomorphic adenoma
    - Malignant
      - Adenoid cystic carcinoma
  - Lymphoid
    - Benign
      - Lymphoproliferative disease
    - Malignant
      - Lymphoma

Inflammatory
  - Infectious
    - Viral
    - Bacterial
  - Noninfectious

Which bug is the most common cause of bacterial dacryoadenitis?
Lacrimal Gland Enlargement

Enlarged lacrimal gland

**Neoplastic**
- Epithelial
  - Benign
    - Pleomorphic adenoma
  - Malignant
    - Adenoid cystic carcinoma
- Lymphoid
  - Benign
    - Benign lymphoproliferative disease
  - Malignant

**Inflammatory**
- Infectious
  - Viral
  - Bacterial
- Noninfectious

*Which bug is the most common cause of bacterial dacryoadenitis?*  
**Bartonella**
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign
- Pleomorphic adenoma

Malignant
- Adenoid cystic carcinoma

Lymphoid

Benign
- Benign lymphoproliferative disease

Malignant
- Lymphoma

Inflammatory

Infectious

Viral

Bacterial

Fungal

Noninfectious

Three common etiologies
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Malignant

Adenoid cystic carcinoma

Two broad categories in the differential

Lymphoid

Benign

lymphoproliferative disease

Malignant

Lymphoma

Inflammatory

Two broad categories in the differential

Infectious

Viral

Bacterial

Fungal

Noninfectious

Pseudotumor

Sarcoid

Thyroid eye dz
Regarding lacrimal gland enlargement, the very general rule of thumb is this:

--If the enlargement occurs in an inflammatory context, think orbital pseudotumor first.

--If the enlargement occurs in a noninflammatory context, think lymphoproliferative disease first.
Lacrimal Gland Enlargement

Regarding lacrimal gland enlargement, the very general rule of thumb is this:
--If the enlargement occurs in an inflammatory context, think orbital pseudotumor first.
Regarding lacrimal gland enlargement, the very general rule of thumb is this:
--If the enlargement occurs in an inflammatory context, think orbital pseudotumor first.
--If the enlargement occurs in a noninflammatory context, think two other words first.
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign
  Pleomorphic adenoma

Malignant
  Adenoid cystic carcinoma

Lymphoid

Benign lymphoproliferative disease

Malignant Lymphoma

Inflammatory

Infectious

Viral

Bacterial

Fungal

Noninfectious

Pseudotumor

Sarcoid

Thyroid eye dz

Regarding lacrimal gland enlargement, the very general rule of thumb is this:
--If the enlargement occurs in an inflammatory context, think orbital pseudotumor first.
--If the enlargement occurs in a noninflammatory context, think lymphoproliferative disease first.
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial
- Benign
  - Pleomorphic adenoma
- Malignant
  - Adenoid cystic carcinoma

Lymphoid
- Benign
  - Benign lymphoproliferative disease
- Malignant
  - Lymphoma

Inflammatory

Infectious
- Viral

Noninfectious
- Pseudotumor
- Sarcoid
- Bacterial
- Fungal
- Thyroid eye dz

If a pt presents with bilateral and symmetrical lacrimal gland enlargement, which condition is the most likely cause?
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign
Pleomorphic adenoma
Malignant
Adenoid cystic carcinoma

Lymphoid

Benign
lymphoproliferative disease
Malignant
Lymphoma

Inflammatory

Infectious

Viral

Bacterial

Fungal

Noninfectious

Pseudotumor
Sarcoid
Thyroid eye dz

If a pt presents with bilateral and symmetrical lacrimal gland enlargement, which condition is the most likely cause? Sarcoid

(Sarcoid-induced lac gland enlargement gets its own set of slides...)
Sarcoid patients

% have ophthalmic involvement

% have no ophthalmic involvement
Sarcoid patients

Half have ophthalmic involvement

Half have no ophthalmic involvement
Sarcoid patients

Half have ophthalmic involvement

Half have no ophthalmic involvement

% have lacrimal gland involvement

% have no lacrimal gland involvement
Sarcoid patients

Half have ophthalmic involvement

Half have lacrimal gland involvement

Half have no ophthalmic involvement

Half have no lacrimal gland involvement
Sarcoid patients

Half have ophthalmic involvement

- Half have lacrimal gland involvement
- Half have no lacrimal gland involvement

Half have no ophthalmic involvement

Enlargement of the lacrimal and salivary glands + keratoconjunctivitis sicca = Mikulicz syndrome
Sarcoid patients

Half have ophthalmic involvement

Half have lacrimal gland involvement

Half have no ophthalmic involvement

Half have no lacrimal gland involvement

Enlargement of the lacrimal and salivary glands + keratoconjunctivitis sicca = **Mikulicz syndrome**
Sarcoid patients

- Half have ophthalmic involvement
- Half have no ophthalmic involvement

For what population is the incidence of ophthalmic involvement far higher—around 90%?

- Very young (<4 years old) children, in whom it is known as early-onset sarcoid.

Early-onset sarcoid presents classically with a triad of signs/symptoms:

- Uveitis
- Arthritis
- Rash
Sarcoid patients

Half have ophthalmic involvement

Half have no ophthalmic involvement

For what population is the incidence of ophthalmic involvement far higher—around 90%? Very young (<4 years old) children, in whom it is known as early-onset sarcoid.
Sarcoid patients

For what population is the incidence of ophthalmic involvement far higher--around 90%? Very young (<4 years old) children, in whom it is known as early-onset sarcoid.

Early-onset sarcoid presents classically with a triad of signs/symptoms—what are they?

- Uveitis
- Arthritis
- Rash
Sarcoid patients

- Half have ophthalmic involvement
- Half have no ophthalmic involvement

For what population is the incidence of ophthalmic involvement far higher—around 90%? Very young (<4 years old) children, in whom it is known as early-onset sarcoid.

Early-onset sarcoid presents classically with a triad of signs/symptoms—what are they?

--Uveitis
--Arthritis
--Rash
Sarcoid patients

Half have ophthalmic involvement

Half have no ophthalmic involvement

For what population is the incidence of ophthalmic involvement far higher—around 90%? Very young (<4 years old) children, in whom it is known as early-onset sarcoid.

Early-onset sarcoid presents classically with a triad of signs/symptoms—what are they?
--Uveitis
--Arthritis
--Rash

Early-onset sarcoid is difficult to distinguish from what other immunologic dz of childhood?
Sarcoid patients

Half have ophthalmic involvement

For what population is the incidence of ophthalmic involvement far higher--around 90%? Very young (<4 years old) children, in whom it is known as early-onset sarcoid.

Early-onset sarcoid presents classically with a triad of signs/symptoms—what are they?
--Uveitis
--Arthritis
--Rash

Early-onset sarcoid is difficult to distinguish from what other immunologic dz of childhood? JRA--juvenile rheumatoid arthritis (aka juvenile idiopathic arthritis, JIA)