Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Hint: NOT ‘benign’ and ‘malignant’—think more broadly than that!
Lacrimal 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

Two broad categories in the differential
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Lymphoid

Two broad categories in the differential

Inflammatory

Two broad categories in the differential

Two broad categories in the differential
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Benign (Finally) Malignant

Two broad categories in the differential

Lymphoid

Inflammatory

Two broad categories in the differential

Two broad categories in the differential

Finally!
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign

Malignant

Inflammatory

Two broad categories in the differential

Lymphoid

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

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

Adenoid cystic carcinoma: Swiss cheese appearance
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Malignant

Adenoid cystic carcinoma

Inflammatory

Lymphoid

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?
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

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Pleomorphic adenoma

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Adenoid cystic carcinoma

Inflammatory

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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

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Malignant

Adenoid cystic carcinoma

Inflammatory

Lymphoid

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?
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

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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?
Basaloid
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).
Two broad categories in the differential

Enlarged lacrimal gland

Neoplastic

Epithelial

Pleomorphic adenoma

Benign

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

Lymphoid

Benign lymphoproliferative disease

Malignant

Enlarged lacrimal gland

Inflammatory

Malignant neoplastic inflammatory epithelial lymphoid adenoma

Benign pleomorphic adenoma

Adenoid cystic carcinoma

Two broad categories in the differential

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

No! The tumor should be removed en bloque, without violating its pseudocapsule.

Why must a pleomorphic adenoma be managed in this way?

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No! The tumor should be removed en bloc, without violating its pseudocapsule.

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

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign

Malignant

Pleomorphic adenoma

Adenoid cystic carcinoma

Benign

Malignant

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

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

Malignant

Adenoid cystic carcinoma

Lymphoid

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

NEVER biopsy to make dx

Malignant

Adenoid cystic carcinoma

Biopsy to make dx

Lymphoid

Inflammatory

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:

Adenoid cystic carcinoma:
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign
  - Pleomorphic adenoma
  - Adenoid cystic carcinoma

Malignant
  - Pleomorphic adenoma: ‘Well circumscribed’
  - Adenoid cystic carcinoma: ‘Bony involvement’

Inflammatory

Lymphoid

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:

NEVER biopsy to make dx

Biopsy to make dx
Lacrimal Gland Enlargement

Adenoid cystic: Bony involvement
(invading the orbital wall, arrow)

Pleomorphic adenoma: Well-circumscribed

Adenoid cystic carcinoma vs pleomorphic adenoma: Imaging
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Neoplastic

Epithelial

Benign

Pleomorphic adenoma

NEVER biopsy to make dx

Malignant

Adenoid cystic carcinoma

Biopsy to make dx

Lymphoid

Two broad categories in the differential

Inflammatory

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?)
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Benign

Malignant

Pleomorphic adenoma

Adenoid cystic carcinoma

Lymphoid

Inflammatory

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’

NEVER biopsy to make dx

Biopsy to make dx
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Two broad categories in the differential

Benign

Pleomorphic adenoma

Malignant

Adenoid cystic carcinoma

Lymphoid

Two broad categories in the differential

Next…
Lacrimial Gland Enlargement

Enlarged lacrimal gland

Neoplastic

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

Lymphoid
- Benign
- Malignant

Inflammatory
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Epithelial

Two broad categories in the differential

Benign

Malignant

Pleomorphic adenoma

Adenoid cystic carcinoma

Lymphoid

Two broad categories in the differential

Benign

Malignant

?

?

Most common within each category?
Lacrical Gland Enlargement

Enlarged lacrical gland

Neoplastic

Epithelial

Benign
- Pleomorphic adenoma

Malignant
- Adenoid cystic carcinoma

Lymphoid

Benign
- 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
- Adenoid cystic carcinoma

Malignant

Lymphoid

Benign
- Benign lymphoproliferative disease

Malignant
- Lymphoma

Inflammatory

Two broad categories in the differential

Next...
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

Inflammatory

Infectious

Noninfectious

Two broad categories in the differential
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

Noninfectious

Types of infections, not specific bugs

Two broad categories in the differential

?
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

Noninfectious

Bacterial
Fungal
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

Which virus is the most common cause of acute dacryoadenitis?
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

Inflammatory

Infecetous

Viral

Noninfectious

Which virus is the most common cause of acute dacryoadenitis?
Epstein-Barr virus
Lacrimal Gland Enlargement

Enlarged lacrimal gland

Two broad categories in the differential

Neoplastic

Two broad categories in the differential

Epithelial

Benign
- Pleomorphic adenoma
- Adenoid cystic carcinoma

Malignant
- Adenoid cystic carcinoma

Lymphoid

Two broad categories in the differential

Benign
- Lymphoproliferative disease

Malignant
- Lymphoma

Inflammatory

Two broad categories in the differential

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
  - Lymphoproliferative disease
- Malignant
  - Lymphoma

Inflamatory

Infectious
- Viral

Noninfectious
- Bacterial

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
  - Lymphoproliferative disease
- Malignant
  - Lymphoma

Inflammatory

Infectious
- Viral
- Bacterial
- Fungal

Noninfectious

Benign
- Lymphoma

Malignant
- Lymphoma

Three common etiologies
Lacrimal Gland Enlargement

Enlarged lacrimal gland

- Neoplastic
  - Two broad categories in the differential
  - Epithelial
    - Benign
      - Pleomorphic adenoma
    - Malignant
      - Adenoid cystic carcinoma
  - Lymphoid
    - Benign
      - 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.
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
    - 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 two other words 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 lymphoproliferative disease first.
If a pt presents with **bilateral** and **symmetrical** lacrimal gland enlargement, which condition is the most likely cause?
If a pt presents with bilateral and symmetrical lacrimal gland enlargement, which condition is the most likely cause? Sarcoid
Lacrimal Gland Enlargement

Bilateral lacrimal gland enlargement in sarcoid
Lacrimal Gland Enlargement

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

% have lacrimal gland involvement

% have no lacrimal gland involvement

Half have no ophthalmic involvement
Sarcoid patients

Half have ophthalmic involvement

Half have no ophthalmic involvement

Half have lacrimal gland 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 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 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?

JRA--juvenile rheumatoid arthritis (aka juvenile idiopathic arthritis, or JIA)
Sarcoid patients

Half have ophthalmic involvement
Half have no ophthalmic involvement

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- --
- --
- --
Sarcoid patients

Half have ophthalmic involvement → Half have no ophthalmic involvement

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-- Arthritis
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Sarcoid patients

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*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)*