Oculoplastic Tumors and Masqueraders

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No financial disclosures
“I am not what I am”

Iago from Shakespeare’s Othello
Case 1

- Healthy 18 mo old girl presented to ER w/ acute L periorbital edema and chemosis, noticeably fussy
- Hx trouble ambulating
- Dx w/ strabismus 2 wks prior elsewhere
- Afebrile, nml WBC, no Hx fever/chills
Exam

- Unable to F&F
- Pupils poorly reactive
- Large ET
- Bil prominent leukocoria (no FHx Rb, not cong, no dog exposure)
- Neither retina could be visualized due to large intraocular masses

Benign or Malignant??
- Admitted
- Head/orbit CT/MRI
- EUA
- Bc high suspicion inflammatory, rather than infectious, cellulitis secondary to advanced Rb, IV steroids initiated
- Rapid resolution of periorbital signs 24 hrs later, discharged home for outpt systemic wu & treatment
- Bc of extensive Dz including neovascular glauc, pt had bil enucleations as more conservative Rx could not salvage globes.
- Path: largely necrotic Rb
- Remains free of Dz 27 mos after surgery
Teaching point

- Rb can present in various ways besides leukocoria
- When presenting as an orbital cellulitis prompt steroids will resolve inflammatory signs and allow for careful exam, imaging, and treatment


Shinder R, Ramirez SA, Chévez-Barrios P, Gombos DS. Retinoblastoma Associated Orbital Cellulitis. ASOPRS Fall Symposium 2011, Orlando, FL
Case 2

- 5 boy brought to ER by mom for unwitnessed fall from bed
- DC home w Dx periorbital contusion/hematoma w/o imaging
- After 1 mo of no resolution pediatrician visit: Dx slowly resolving hematoma
- After 2 mos of no resolution back to ER -> 1st ophtho consult
Benign or Malignant??
Pre Op Labs

Hb: 10
Platelets: 100k
WBC: 5
Surgical Findings:

- tumor, gross bony lytic change of lateral orbital wall, heavy bleeding

Frozen Section – inconclusive

Extended lid crease incision was done and orbital biopsy was taken.

Frozen Section – inconclusive, but favoring Rhabdomyosarcoma
Two drains placed removed POD#2. Patient admitted to PICU

Post Op Labs

Hb: 4
Platelets: 20k

Transfusion

2 units RBC’s
1 unit of platelets
Final Pathology:
Metastatic Neuroblastoma
- Systemic wu: widespread osseous mets to skull base & calvarium, BM
- Primary: 3.3 x 2.5 x 3.5 cm right superior mediastinal paraspinal mass
Treatment thus far....

- Several cycles chemo
- Right posterior thoractomy tumor excision
- RT

- Repeat scans show bone and orbit lesions markedly diminished, no recurrence at mediastinum
Teaching point

- Mets neuroblastoma can present acutely similar to trauma, child abuse
- Must suspect in any young child with acute orbital signs/symptoms
- Timely Bx, metastatic wu, multidisciplinary team recruitment are key
Case 3

- 70 M underwent excision SCC of R cheek 2 yrs prior by dermatologist w/ pos margins
- Developed R VII palsy w/ RLL paralytic ectropion few days after excision
- Dx w/ Bell’s palsy & had ectropion repair by oculoplastic surgeon
- Ectropion recurred after 8 mos
- MRI ordered & referred for eval
- Path of original lesion confirmed SCC
- No perineural invasion noted in specimen
- Pt denied pain or paresthesias
Exam

- Va OD – 20/400 OS – 20/20
- Hypesthesia R V1, V2
- R peripheral VII palsy
- No LN’s
Benign or Malignant??
Dx of recurrent SCC of the cheek w/ extensive perineural spread & resultant multiple cranial neuropathies was made
- Systemic w/u neg
- Rad onc & med onc consults
- Given extensive skull base spread, felt that tumor was not surgically resectable
- Since pt had no pain, RT deferred
- IV chemo (carboplatin & paclitaxel) was given
- Antibiox gtts & oint
- RUL gold wt, LTS, lat tars
After 2 cycles of chemo, pt had significant resolution of clinical & radiographic signs of perineural spread

20/50 OD

Resolved K ulcer

Residual but improved V1/V2, VI, VII palsy
3.5 yrs after chemo - no clinical or radiographic evidence of recurrence = durable response

We are all to this day surprised with this great response of perineural SCC to chemo!
Teaching point

- Bell’s palsy is only Bell’s palsy if you have ruled out other causes of VII palsy (Dx of exclusion)
- SCC has predilection for perineural spread (V, VII most common), & this typically carries a poor prognosis
- Combination of V & VII palsy very dangerous for the cornea

56 M presented w painless L proptosis, hypoglobus, & diplopia for several months
Evaluated at outside institution: found to be euthyroid, and CT orbits obtained
Patient was diagnosed with TED and referred for oculoplastic evaluation
Our exam:

- Va 20/20 OU
- mild limitation in supra-, infra-, and adduction, normal abduction OS
- OS proptosis, hypoglobus confirmed
Benign or Malignant??
TED?

Few findings were atypical for TED:

- Euthyroid
- Hypoglobus
- Unilateral presentation
- Lack of lid retraction
- Normal abduction w limited adduction
- Male
We recommended EOM biopsy - pt declined, wished to continue with observation

10 months later:
- Va OS declined to CF
- Proptosis progressed
- Ductions more severely restricted
- Now had anterior orbital mass visible in the superior fornix
Patient now agreed to intervention, and an incisional biopsy confirmed a diagnosis of:

- Liposarcoma
- Systemic w/u neg for mets
- Patient underwent L exent
- 60Gy of RT
- No recurrence and is free of disease at last follow up 66 months following surgery
Teaching point

- Orbital malignancy can mimic TED due to overlap of clinical and radiographic findings, including proptosis, multiple EOM enlargement, and restrictive ophthalmopathy w diplopia

- Other etiologies should be considered when presentation is atypical (primary or metastatic CA), Bx when warranted

Conclusion

- Malignancy can at times masquerade as a more benign entity, vice versa
- A high degree of suspicion is paramount to timely diagnosis and treatment
Thanks