Grand Rounds Case Presentation

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Preceptors: Dr. Singh, Dr. Palestine, Dr. Seibold
Case Report

• 3 month old male presented to CHC with 3 weeks of irritability, right-sided epiphora, photophobia and cloudy cornea
• Vision
  – Right: Blink to light
  – Left: Fix and follow
• Pupils
  – Hazy view right, no rAPD
• IOP (Icare) - *difficult*
  – Right: 63
  – Left: 15
• Plan: Started on topical timolol, dorzolamide, latanoprost right eye, scheduled for EUA
EUA the next day

- IOP (tonopen)
  - Right: 43
  - Left: 11

- Horizontal Corneal Diameter
  - Right: 12.5
  - Left: 12.0
Slit Lamp Exam
Examination

• SLE:
  – L/L: wnl OU
  – C/S: 1+ injx OD, wnl OS
  – K: 1+ edema, no Haab’s striae OD, wnl OS
  – AC: 1-2+cell, vascularized mass appeared to arise from the inferotemporal angle in OD, wnl OS
  – I: immature iris vessels OD, wnl OS
  – L: clear OU

• DFE:
  – Right: c/d 0.65, macula/vessels/periphery wnl
  – Left: c/d 0.3, macula/vessels/periphery wnl

• Gonio: Immature angle structures OD with inferotemporal mass obscuring view to angle. Normal OS.
Case Summary

- 3 month old male
  - Angle mass with vascularization
  - AC inflammation
  - Immature iris vessels
  - Glaucoma (High IOP + cupping)
  - Normal DFE and SDE
Differential Diagnosis?
Differential Diagnosis

- Juvenile Xanthogranuloma
- Leukemic infiltrate with pseudohypopyon
- Retinoblastoma
- Ciliary body medulloepithelioma
Further Testing?
Further Testing?

• Complete physical exam
• UBM
• Anterior segment angiography
• AC paracentesis
Complete Physical Exam

- No cutaneous lesions identified during complete exam during EUA
- No cutaneous lesions identified on dermatology consult
UBM

OD SCAN: 82
TVGain: 0
Volts: 127
Orient: 6 o'clock

Probe: 50MHz
Sound: 1550 m/s
Angle: 35 deg
Anterior Segment FA
AC Paracentesis

• On entering the AC, multiple points of the iris vasculature hemorrhaged
• AC washout required
• Cryotherapy applied to corneal wounds to prevent potential seeding of malignant cells

• Pathology: Hemorrhage and inflammatory cells which were predominantly macrophages and monocytes without evidence of malignant cells
Juvenile Xanthogranuloma

- Classically described as self limited non-langerhaans cell histocytic cutaneous disorder affecting infants and young children
- Epidemiology
  - Slight male predilection, no sex bias for ocular involvement
  - Present at birth in 10%
  - 85% of skin lesions present by 1 year of age
- Skin lesions are typically:
  - firm, slightly raised papulonodules several millimeters in diameter.
  - tan-orange in color
  - frequently on the head and neck, but many extracutaneous sites have been reported.
- Extracutaneous lesions reported in nearly every organ system
Pathophysiology

- Cause unclear
- Thought to be abnormal macrophage response to tissue injury resulting in granulomatous inflammation
- Pathology: dense histiocytic infiltrate with large vacuoles, Touton giant cells in 85% of cases
Ocular JXG

- The eye is the most common extracutaneous location for JXG to manifest
- 92% of pts less than 2 years of age
- Ocular manifestations occur in 0.4-10% of pts with JXG
  - Recurrent hyphema
  - Iris mass
  - Uveitis
  - Heterochromia
  - Corneoscleral, conjunctival, and posterior segment lesions have also been reported
- Glaucoma may occur as a secondary complication in as many as 75% of eyes with ocular JXG
- 50% of pts with ocular JXG have skin lesions
- 50% of pts with ocular JXG present initially to ophthalmologist
Glaucoma in JXG

- Reported in up to 75% of eyes with ocular JXG
- A significant cause of visual morbidity in JXG
- Likely multifactorial
  - Inflammatory cells shed from the iris +/- monocyte infiltrate in the TM
  - Red blood cells from recurrent hyphema impair trabecular outflow
  - Direct angle obstruction by iris mass may lead to increased intraocular pressure.
JXG Diagnosis

• Typically a clinical diagnosis
• Bx of skin or ocular surface lesions if present and dx unclear
• Other tests can help support the dx:
  – AC tap: Typically demonstrates histiocytes, monocytes
  – UBM: Variable location (iris stroma, angle, etc), homogenous internal reflectivity, absence of vascular channels
  – Iris FA: Early hyperfluorescence of lesion, diffuse late iris leakage
JXG Treatment

- Cutaneous JXG typically resolves spontaneously and does not require treatment
- Ocular JXG should be treated
  - Control inflammation
    - Topical steroids typically do not control ocular lesions
    - Local (i.e. subconjunctival or subtenons) steroids
    - Systemic steroids
    - Systemic immune suppression
  - Control IOP
    - Topical pressure lowering gtts
    - Systemic CAIs
  - Shrink the lesions
    - Radiation therapy
    - Intracameral avastin
    - Surgical resection
Our Patient

• Received SubTenons Kenalog at time of EUA/UBM/FA
  – Persistent inflammation and recurrent hyphema causing IOP spikes despite topical and systemic IOP lowering
• Repeat SubTenons Kenalog 4 weeks later
  – Still with multiple rebleeds and IOP spikes despite max gtt tx, PO diamox and PO prednisone
  – Angle mass smaller but still present
• Pediatric Ahmed placed 7 weeks after presentation
  – IOP controlled but developed cataract
  – Angle mass regressed by time of surgery
Our Patient

• Cataract extraction 4 months after presentation
  – Left aphakic and aphakic contact lens placed
  – Persistent inflammation despite topical and systemic steroids

• Started on MTX PO 5mg weekly
  – IOP controlled off gtts
  – Inflammation controlled on MTX
  – Continuing treatment for amblyopia
Questions/Comments?
References