Complicated Cataract to Intraocular Tumors, Beware of the unexpected

Ihab Saad Othman, MD, FRCS
Professor of Ophthalmology
Cairo University

• In this part of the world: We Master Phakoemulsification
Intraoperative/Second day postoperative

- Following cataract surgery:
  - Surprise:
    - Ciliary body mass !!!
  - DD:
    - Suprachoroidal hemorrhage
    - Mass = Tumor

Anatomy

- **Lens:**
  - Elastic, avascular, transparent highly refractile biconvex disc.
  - Lying in posterior chamber

- **Ciliary body:**
  - Triangular structure
  - LOCATED IN A SILENT AREA OF THE EYE
Materials and Methods

• Retrospective chart review:
  - 465 cases of uveal mass
  - 560 cases of pediatric IO tumors
  - Managed from September 2000 to December 2016
  - Identification of anterior located tumors compressing the lens

Results:

• We identified 79 cases of CB masses compressing the lens and inducing:
  - High astigmatism
  - Focal cataract
  - Operated from cataract/glaucoma surgery
Adult cases

- 55 YOF
- Focal cataract
- CB and choroidal infiltration identified
- Managed elsewhere by enucleation

- Extensive uveal and vitreal involvement
• Non-Hodgkins lymphoma

• Typical management:
  - FNAB
  - Choroidal biopsy
  - Chemotherapy
  - External beam radiotherapy
Beware of new onset astigmatism/Focal cataract

47 YOM, Phako IOL 1 year prior
58 YOF, Focal cataract

- 55 YOF
- **History:**
  - Cataract extraction 1 year ago
- **CP:**
  - Epibulbar mass
  - Progressive enlargement
• **Modified enucleation**

• **1 year follow up:**
  - Liver metastases

• **72 YOF**

• **History:**
  - Cataract extraction and IOL 8 months ago
  - Growing epibulbar mass
  - Huge sentinel vessels
  - Follow up for 8 months !!!!!!!!
• **Management:**
  - Modified exenteration
  - Spindle cell melanoma
  - Surgical margins are free

• **CAUTIOUS FOLLOW UP**

• 44-year-old male
• DVA OD OS: 6/6 Normal
• Past History:
  - Cataract extraction OD 8 years ago
  - SBP OD 6 years ago
  - Recurrent RD \(\rightarrow\) PPV, endolaser 4 years ago
  - Recurrent attacks of unexplained uveitis and 2ry glaucoma \(\rightarrow\) SST
• Systemically healthy
• **VA OD: PL BP**

• **Anterior segment:**
  - Malignant hypopyon
  - Granulomatous KPs
  - Markedly distorted iris
  - Aphakia

• **Fundus:**
  - Not seen

• **MRI:**
  - **T1: Sagittal:**
  - Soft tissue mass occupying the anterior portion of the globe in CB region
  - Aphakia
  - Normal ON stump
- **MRI**
  - Coronal T1 Gd
    - Enhancing soft tissue mass in CB region
    - No EOE

- **MRI**
  - Axial, T1, Gd
    - Mildly hyperintense enhancing soft tissue mass in CB region
• **MRI:**
  - Axial MRI T2
    - Hypointense soft tissue mass in CB region
    - Hour glass globe

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**Management**
- **Histopathology:**
  - Adult Medulloepithelioma:
    - Non-teratoid
    - No malignancy
    - AC invasion
    - Retinal invasion
  - Explanation:
    - Toxic cataract vs focal cataract
    - Exudative RD before buckle
    - Rhegmatogenous element following SBP, before PPV
    - No masses in CB region during PPV
    - Toxic uveitis and secondary glaucoma
    - Mass effect

- **Medulloepithelioma:**
  - Tumor of NPCE
  - Verhoeff: 1904 Teratoneuroma
  - Fuchs: 1908 Diktyoma(Net-like)
  - Ginker: 1931 Medulloepithelioma (primary medullary epithelium)
• **Spread:**
  - Adult type: very rare: due to EOE
    - Optic nerve extension
    - Lymphatic extension
  • **Management:**
    - Enucleation
    - Local resection (watch for recurrences)
      - Radiotherapy
      - Chemotherapy

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**Young age group**

• 8 YOF
• Focal cataract
• Irregular astigmatism
  - Medulloepithelioma
• Brachytherapy

• Lesion stable over 4 years of follow up

• 1.5 YOM
• Iris, CB mass
• Marked astigmatism
• Minimal focal cataract:
  - Scheduled for enucleation
• **DD:**
  - Langerhan’s cell histiocytosis (JXG)
  - Rhabdomyosarcoma
  - Granulomatous lesions
  - Others

• **Plan:**
  - FNAB:
    • Epithelial cells
    • No malignancy

• **Management:**
  - Lamellar Iridocyclectomy
• Pathology:
  - Ectopic lacrimal gland

Retinoblastoma cases

• Presence of cataract is against the diagnosis of retinoblastoma:

• This is not an absolute statement
Mohamed Hefny

- 8 year-old boy
- History:
  - Multiple surgeries
  - Cataract extraction
  - PPV
  - Multiple iris biopsies
  - Despite presence of round blue cell tumor in iris !!!!!!

Anterior Segment seeds are misleaders:

- Older age at presentation
- Flat retinal pattern
- Good visual acuity

ihabsaad@hotmail.com
• 6 YOF
• Cataract surgery 9 months ago
• Epibulbar mass
• Refusal of enucleation for 3 months
‘Mystery Case’

Ihab Saad Othman, MD, FRCS
Professor of Ophthalmology
Cairo University

ihabsaad@hotmail.com

History

• 62 year old caucasian woman

• Recurrent anterior uveitis OD

• Type II Diabetes

• Referred re ? Ciliary body melanoma

ihabsaad@hotmail.com
Examination

• 6/12 OD  6/7.5 OS

• Anterior synechiae OD

• No active intraocular inflammation

• Normal IOP

• Mild nonproliferative diabetic retinopathy OU
UBM - anteroposterior
Differential?

- Adenoma (non-pigmented ciliary epithelium)
- Medulloepithelioma
- Metastasis
- Melanoma
• Granulomatous inflammation

• Non-caseating

• ZN-negative

• ?? Sarcoidosis, chest X-ray normal, ACE normal

• Extensive endophytic retinoblastoma
• Massive optic nerve and extraocular extension
• Prior management
  - Lensectomy, SST, PPV
• Further management:
  - Chemotherapy x 9 cycles
  - EBRT
  - Intrathecal MX
Clues U should not Miss

- Recent Irregular astigmatism:
- Sentinel vessels

- Epibulbar pigmentation
- Epibulbar mass
• Angle infiltration

• Transillumination:
  - A forgotten art
• **Ultrasonography:**
  - Emphasis on CB region

• **UBM**
  - Subtle CB masses
• **Scanning images:**
  - CT
  - MRI

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**Discussion**

• *General Ophthalmologist and cataract surgeons should be aware of this presentation despite being rare*
• *Simple diagnostic tests as in-office fundus examination, transillumination, US would clinch diagnosis*
• *Most operated eyes are deemed to enucleation*
• *Early diagnosis and timely intervention is imperative in SURVIVAL*
• Remember:
  - WHEN DEALING WITH CANCER
    YOU ARE DEALING WITH LETHAL
    DISEASE
    YOU ARE DEALING WITH HUMAN LIFE

Thank YOU