From a lamb to a beast
A scary tale

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History

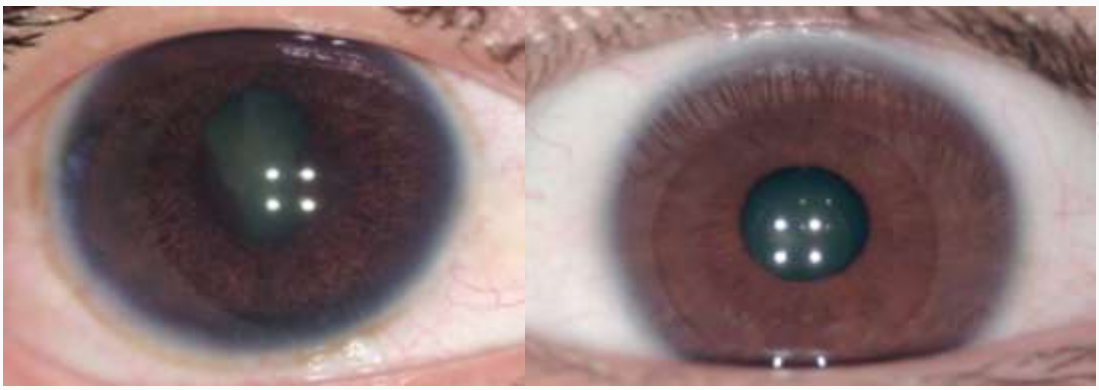
- 42 years old female.
- Housewife.
- Medically free.
- No significant past history of ocular trauma or surgery.
- Complaints of gradually progressive diminution of vision in her right eye over the past 6 months.

On examination

	OD	OS
BCVA	**6/60**	6/9
Refraction	-0.75/-1.25x135	-0.25/-0.75x55
EOM	Free	Free
Anterior. Segment	????	Free
Fundus	Hazy view **C/D= 0.7**	Normal Fundus C/D= 0.4
IOP	**20 mmHg**	16 mmHg

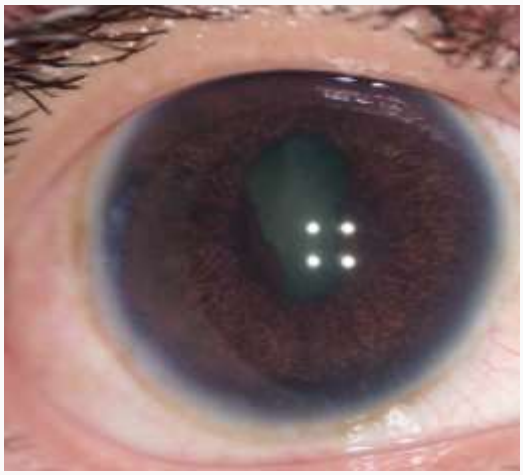
On Examination

Right eye Left eye



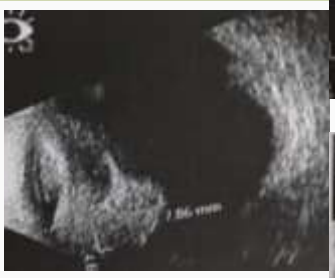
So, what's your differential diagnosis?

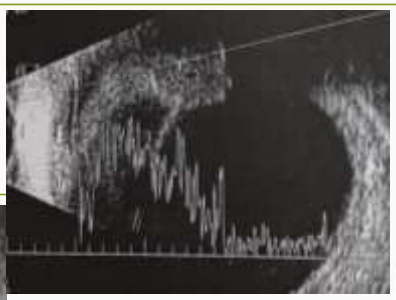
- Iridociliary melanoma.
- Diffuse iris nevus.
- Iris cyst.
  - Iris pigment epithelium cyst.
  - Epithelial inclusion cyst.
- Others → Uveitis.

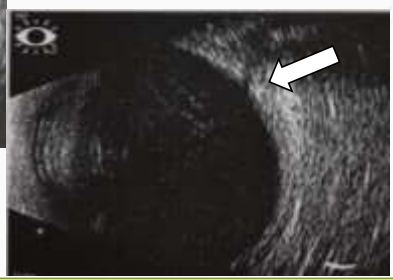


US

- Opaque displaced lens by temporal ciliary body mass.
- Retina in place
- Evidence of large deep optic disc cup +/- 0.7

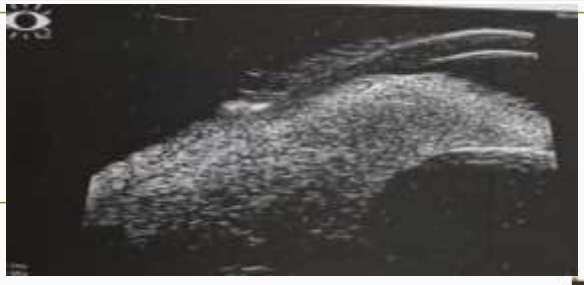


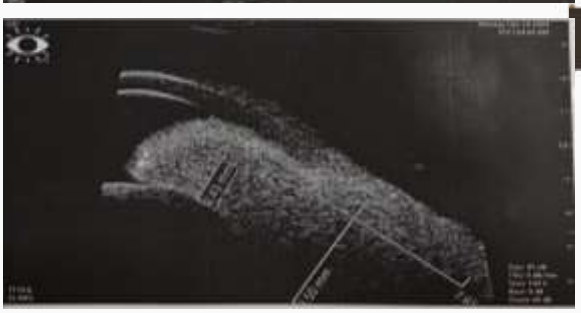




UBM

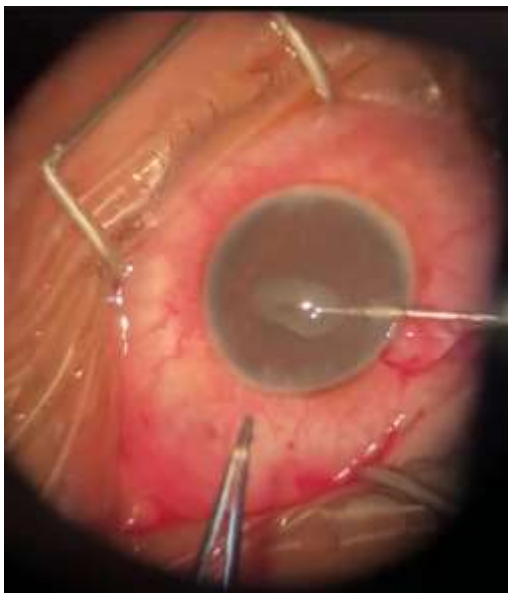
- Iridociliary mass extending temporally from 6 o'clock to just before 12 o'clock.
- Indenting the lens, infiltrating zonules.
- Dimensions:
  - Transversely: **7.6 mm**.
  - Vertically: **2.4 mm** (Iris), **3.5 mm** (CB).
- Occluded superior and temporal angles.
- No extrascleral extension.





So, what's next?

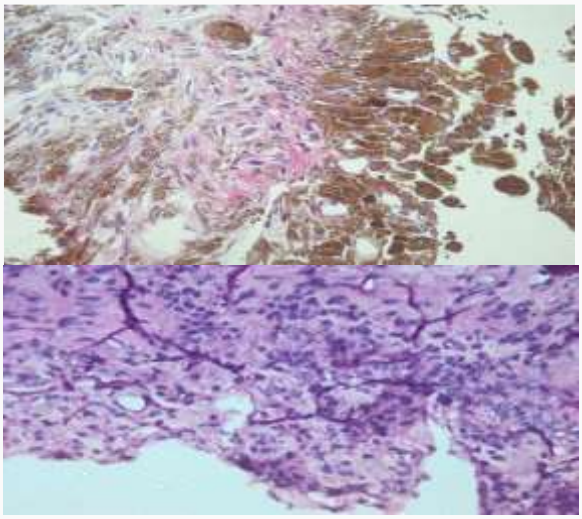
A biopsy was taken from the mass and was sent for histopathology



Histopathology

- Polygonal cells heavily laden with melanin.
- Better viewed after bleaching→ small nuclei with abundant cytoplasm, minimal pleomorphism and no mitotic activity.

Iris melanocytoma



What is a MELANOCYTOMA?

What is a MELANOCYTOMA

- First described by Zimmerman and Garon in 1964, as a variant of melanocytic nevus.
- Cogan suggested **magnocellular nevus** as a better descriptive term.
- Originally described for optic nerve melanocytoma but can develop anywhere in the uveal tract.
- In a series of 2510 cases with iris tumors → only **3%** were melanocytoma.
- Diagnostic challenge→ mimics uveal nevi and malignant melanoma.

What is a MELANOCYTOMA

- Mean age: 36 years but some cases were reported in children.
- More common in females.
- Dark brown or black lesion with corrugated friable surface→ satellites on iris surface (53%), trabecular meshwork(40%).
- Composed of deeply pigmented plump or polygonal cells with abundant cytoplasm and small round uniform nuclei
  - **Type 1 melanocytoma** cells: large polyhedral cells with giant melanosomes (1 to 5 μπι) and few cytoplasmic organelles
  - **Type 2 melanocytoma** cells consisted of small spindle-shaped cells with infolded nuclei, conspicuous nucleoli, small melanosomes (0.3 to 1.0 μπι), and abundant cytoplasmic organelles.

Back to our patient

- Metastatic work up was done → insignificant
  - CT Brain
  - CT Chest
  - CT Abdomen and Pelvis
  - Liver enzymes
- Patient was scheduled for monthly follow up
  - For 6 months→ the condition was stationary but cataract was gradually increasing in density

Until one day

- The patient presented in a follow up with acute significant drop of vision.
- **On examination:**
  - BCVA: **CF 30 cm.**
  - Marked pigment dispersion in anterior chamber.
  - IOP **28 mmHg.**
- She was injected subtenon triamcinolone, prescribed topical steroids and topical antiglaucoma eye drops.
- **After 2 days;**
  - AC was completely quiet and IOP back to 18 mmHg and VA 3/60.

So, what happened?

- Spontaneous necrosis→ pigment dispersion→ free pigment or pigment-laden macrophages accumulate in TM → obstruction of aqueous outflow

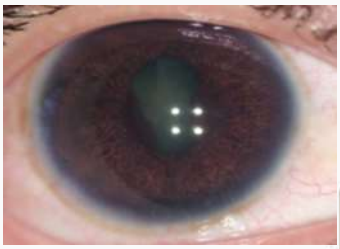
Melanocytomalytic Glaucoma

Melanocytomalytic glaucoma

- Commonly associated with iridociliary melanocytoma.
- Suggested to occur secondary to interruption of intrinsic blood supply with high metabolic activity and friable surface → areas of necrosis with pigment dispersion.
- In a series of 47 cases of iris melanocytoma, elevated IOP in 11% at 5 years and 50% at 15 years
- Other mechanisms for IOP elevation.

In subsequent follow up visits

- The lesion clinically appears to increase in size
- New UBM was requested→ increase in size
  - **Transversely**: **9.7 mm** instead of 7.6 mm
  - **Vertically**: **4 mm** instead of 2.4 mm



Back to literature

- Melanocytoma is a relatively stationary lesion→ growth occurred in 23% of the patients at 5 years and 48% at 10 years.
- Subtle enlargement does not necessarily imply malignant change, but more profound and rapid growth is generally considered a sign of malignant transformation into melanoma.
- Only few cases of iris melanocytoma are reported to have undergone malignant transformation.

PET-CT scan

Findings:

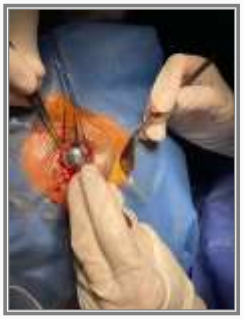
- **Right eye**: mild increased FDG uptake by hyperdense lesion seen in the anterior aspect of temporal side of the eye ball with SUVmax 3.5 and measures about 5.0 x 2.5 mm. The lesion is seen abuts and displacing the lens to the nasal side.
- **Left eye**: no FDG-avid focal lesions.

Opinion

- Mild metabolically active hyperdense lesion in the temporal side of right as described.
- No evidence of other hypermetabolic lesions.

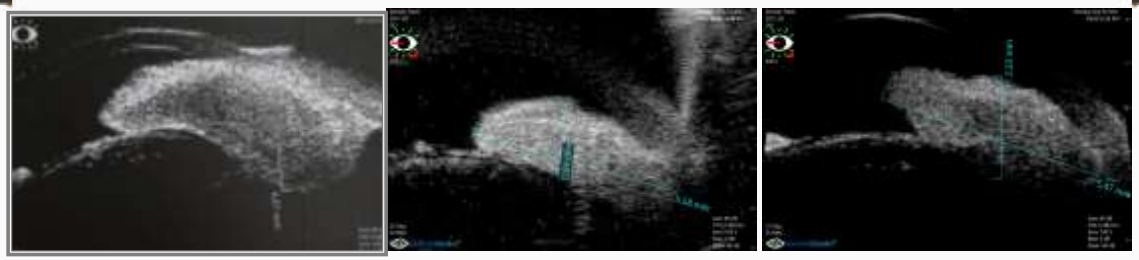
Brachytherapy was decided

- Ruthenium 106 radioactive CIB plaque was used.
- Applied for 6 days to deliver a dose of 110 Gy to tumor apex.
- Lateral rectus was disinserted to allow for proper plaque placement and reinserted after plaque removal.



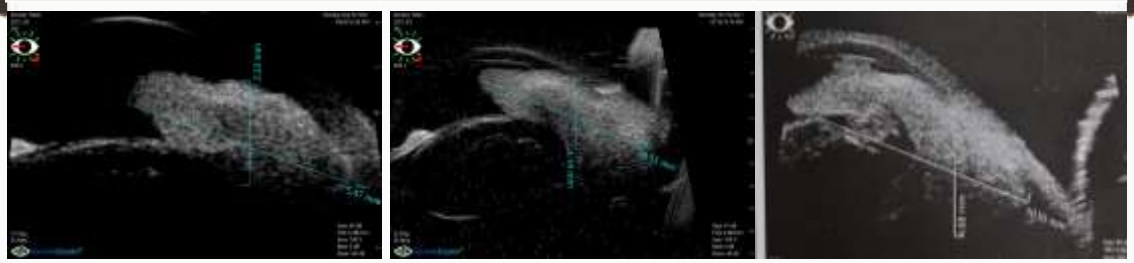
Serial follow up UBM initial marked regression

Before plaque	After 6 weeks	After 3 months
9.7 mm	5.58 mm	5.47 mm
4.07 mm	2.04 mm	2.32 mm



Serial follow up UBM but unfortunately

After 3 months	After 4 months	After 5 months
5.47 mm	8.53 mm	10.55 mm
2.32 mm	3.14 mm	4.68 mm

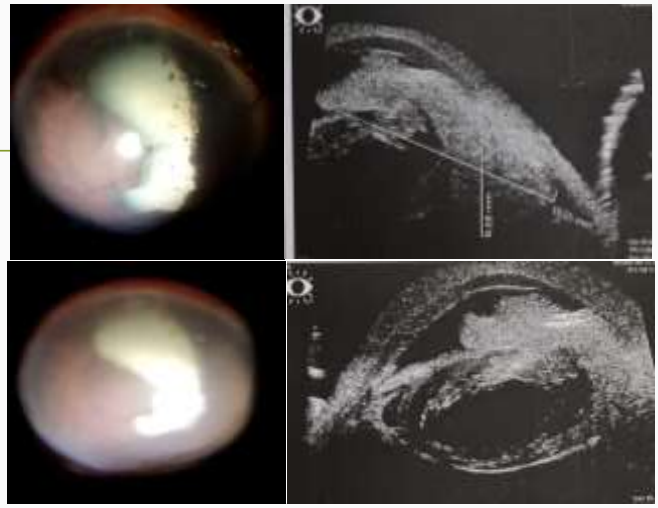


5 months after brachytherapy

- VA: dropped to **HM**
- Mass increased markedly in size
- Total lens opacity with subluxation
- IOP: **30 mmHg**

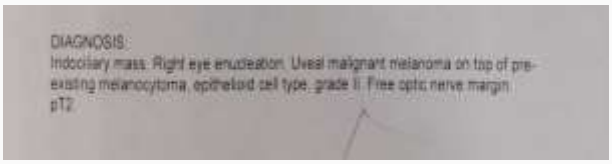
So after counselling the patient;

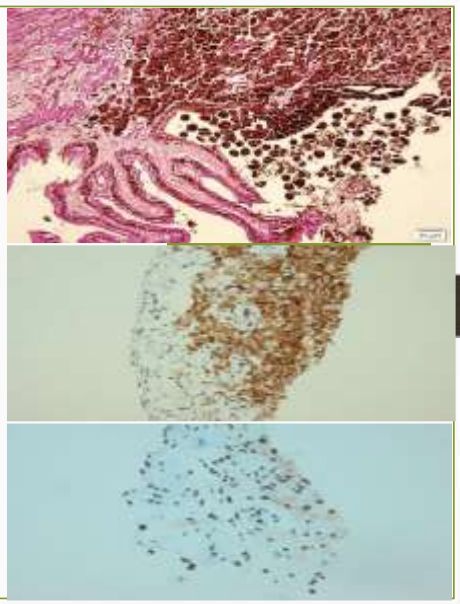
Enucleation of the right eye was done



Histopathology

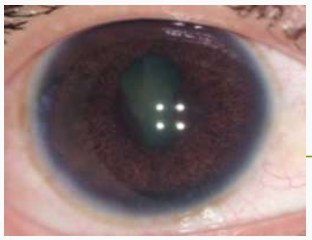
- Malignant epithelioid and polygonal tumor cells with abundant melanin showing nuclear pleomorphism
- Strong membranous **HMB45 staining** indicating melanocytic lesion
- Strong nuclear staining of **Ki67** ndicating a high proliferative index of the tumor cells.



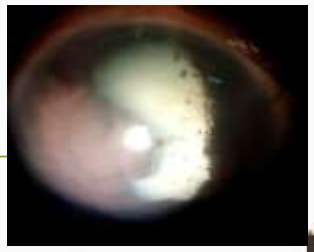


Back to literature
Management of iridociliary melanocytomas

- **Watchful observation** → small stationary lesion.
- **Local resection** → lesions < 3 clock hours.
  - Significant pigment seeds.
  - Uncontrolled elevated IOP.
  - Significant tumor growth or suspected tumor growth.
- **Brachytherapy**
- **Enucleation**
  - Blind painful eye.
  - Malignant transformation.



Take home messages



- Iridociliary melanocytoma is a rare benign tumor → may involve uveal tissue.
- Melanocytomalytic glaucoma, pigment dispersion and seeding are common.
- In spite of being benign, watchful follow up is a must → you don't know when malignant transformation will happen.
- Don't hesitate in taking tough decisions → Enucleation can save a patient's life when other ways fail.

Acknowledgment

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- Special thanks to:

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Thank you ☺