Pigmented Iris Mass: Differentiating Between Melanoma and Nevus

Tessa Plana, OD, Michael Sullivan-Mee, OD, FAAO, Suchitra Katiyar, MPH, OD, Denise Pensyl, OD, MS, FAAO

Abstract
Iridociliary body melanoma is a condition that can be catastrophic to the eye or the patient’s life. This report discusses a case in which ultrasound biomicroscopy is used to help distinguish whether melanoma is present.

I. Case History
- A 32-year-old white male presented for his first eye exam 3 months ago and was diagnosed with an iris nevus at 4 o’clock that was growing into the angle. The patient subsequently referred to our clinic for further workup. The patient reported he had seen the nevus 2-4 years ago but didn’t think it was growing and wasn’t too concerned about it.
- The patient had no other ocular history. Medical history included post-traumatic stress disorder, shoulder instability, overweight, and snoring. The patient's only medication is bupropion 300mg daily.
- No family history of cancer

II. Pertinent findings
- Without correction, Snellen visual acuities were 20/20 in each eye.
- Entrance/biomicroscopy findings:
  - The patient had equal, round and reactive pupils. Extraocular muscles demonstrated full range of movement. The eyes were aligned, and confrontation fields were full in each quadrant of each eye.
  - The patient’s lids, lashes, conjunctiva, cornea, and anterior chamber were unremarkable.
  - There was a dark thick neoplasm of the inferior-nasal iris with a size measured at 3.1x1.2mm. The neoplasm was pushing on iris, elevated into angle, and contacting the base of cornea. The iris of the left eye was unremarkable.
  - Goldman intraocular pressures were 13mmHg OD and 14mmHg OS.
- Gonioscopy:
  - Gonioscopy exam of the right eye revealed the angle open to ciliary body 360 degrees except inferior-nasal where angle was obscured by a pigmented neoplasm. There was mild patchy pigment elsewhere that was not consistent with melanoma infiltration of angle (i.e. no sign of beginning or advanced ring melanoma). The iris had a flat approach. There were no intrinsic vascular irregularities, neovascularization, or hemorrhage.
  - The left eye was also open to ciliary body 360 degrees with mild pigment and a flat iris approach.
- Ultrasound biomicroscopy (UBM) with VuMAX Ultrasound System:
  - UBM revealed localized thickening of the iris and underlying ciliary body in the inferior-nasal portion of the eye. The thickening did not extend beyond where iris thickening was located. There were no obvious cystic components seen on UBM exam.
Measurement of the ciliary body portion of the tumor on UBM was about 3.9 millimeters versus about 2.0 millimeters in the adjacent area of non-thickening.

The left eye was not imaged on UBM.

- Anterior segment slit lamp camera images were taken of the iris mass.
- Dilated fundus exam:
  - The right lens had trace amount of pigment on the anterior lens capsule with no induced cataract, and the left lens was clear.
  - The patient had moderate optic nerve cups in average disc sizes with unremarkable optic nerve rims, macula, posterior pole, vessels, and a flat and intact periphery.
- Laboratory chart review:
  - The patient had recent normal liver labs including total protein, albumin, alkaline phosphatase, aspartate aminotransferase, total bilirubin, direct bilirubin, and alanine aminotransferase.
  - Liver function labs were repeated by an ophthalmologist on a followup visit to investigate liver metastasis, and labs were still normal 2 weeks after our visit.
- Consult with ophthalmologist:
  - During a consult with ophthalmology 2 weeks after our visit, the findings were unchanged from our exam. The ophthalmologist ordered another liver function test and a position emission tomography (PET) scan.
- Radiology studies:
  - The PET scan revealed probable liver hemangioma and some small lung nodules. There were no definite findings of metastatic disease.

III. Differential diagnosis

- Iridociliary body melanoma
  - Factors that increase suspicion for malignancy include: new lesions, large, elevation, irregular borders, disrupted adjacent anatomy, cataract, increased or abnormal vasculature, increased or patchy pigment, high intraocular pressure, astigmatism, sentinel episcleral vessels, corectopia, and hyphema (1).

- Iridociliary body nevus
  - Factors that lead towards a diagnosis of nevus include: congenital or longstanding onset, small, round, normal adjacent anatomy, and flat. In general, melanocytic lesions smaller than 2.5 to 3.0 mm in thickness are less suspicious for being neoplastic (2). Iris lesions are classified as cystic or solid, and cystic are generally benign (3). UBM can be used to help differentiate between nevus and melanoma by better visualization of the border irregularities, base and thickness size, shape, and location. UBM can also better show its inner consistency such as cystic or solid, vascularity, acoustic features, and homogenous or heterogenous pattern (4).

IV. Diagnosis and discussion

- The patient was assessed as having a pigmented iridociliary body neoplasm of the right eye, likely melanoma. The lesion appeared localized without adjacent infiltration of the angle. Consult with an ocular oncologist is pending.
- Iridociliary body melanoma is a neoplasm of the iris and ciliary body. Melanoma is the most common primary intraocular malignancy. The overall survival rate has not significantly
improved since the early 1970s (5). Timely treatment may prevent growth, enucleation, metastasis, or may save the patient’s life. Ultrasound biomicroscopy can be used to better determine the neoplasm’s characteristics. Also, ultrasound is especially helpful in neoplasms of the iris that extend into ciliary body (4).

- Prognosis: Size, location, intermediate or epithelioid cell type, presence of extraocular extension, high mitotic rate, and lymphocytic infiltration all factor in to the prognosis of the condition. (1).
- Risk factors for iris melanoma can be remembered using the ABCDEF guide: A= age young (<40), B= blood in anterior chamber, C= clock hour of mass inferiorly, D= diffuse configuration, E= ectropion uveae, F= feathery margins (3).
- For iris melanoma, at 5 and 10 years the metastasis rate was 4% and 7%, respectively. For ciliary body melanoma, at 5 and 10 years, the metastasis rate was 19% and 33%, respectively (1).
- Metastasis may sometimes be used as a replacement for mortality based on the current assumption that uveal melanoma metastasis ultimately leads to metastatic death (6).
- Metastatic iris tumors are more common from carcinoma of the breast, lung, kidney, and skin (3).

V. Treatment, management

- The management for this case included referral to an ophthalmologist and ocular oncologist. A PET scan was ordered and reviewed to look for metastasis. Liver function labs were ordered and reviewed due to common melanoma metastasis to the liver.
- The ocular oncologist will assess the lesion and its recommended treatment. Current treatment modalities include transpupillary thermotherapy, plaque radiotherapy, charged particle irradiation, local resection such as iridocyclectomy and iridectomy, enucleation, or orbital exenteration (1). Radiotherapy is the most common treatment for lesions that qualify, and large lesions are most often treated with enucleation (5).
- There is better prognosis with iris melanoma compared to ciliary body melanoma. Also, there is better survival in younger patients (1).

Bibliography
VI. Conclusion

- Take away points:
  - Ultrasound biomicroscopy is a vital tool for aiding differentiation between melanoma and nevus of that may extend posterior to the iris.
  - Liver function labs and PET scan can be ordered to rule out metastasis before patients visit an ocular oncologist.
  - Timely diagnosis and referral is indicated for iridociliary body lesions to possibly improve the patient's outcome.