Dispelling Rumors about Tumors

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Case

• 65 year old female presents with flashes
Diagnosis: Choroidal Melanoma

• 5% of all melanomas in the US
• most common primary IO tumor in adults
• 6 cases/million → 1500 cases per year in US
• 50-70 years/women=men/Caucasian
• Diagnosis based on fundoscopy + ultrasound

Rumor #1: Everything that’s pigmented is a melanoma
Rumor #2: Everything that’s pigmented and elevated must be a melanoma
Case: Melanoma v. Nevus?

Melanoma v. Nevus?
Melanoma v. Nevus?

The only melanoma in the bunch
Diagnosis: Choroidal Nevus

- Benign tumors
- Collection of bland spindle A melanocytes
- The edges are defined but not sharply demarcated
- Dark brown or grey pigmentation
- Amelanotic not unusual
- High risk features which predispose to growth
- Growth may or may not be a sign of malignancy

Which nevi grow?

<table>
<thead>
<tr>
<th>Feature</th>
<th>Feature in Nevi that progress to Melanoma (%)</th>
<th>HR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thickness &gt; 2mm</td>
<td>19</td>
<td>2</td>
</tr>
<tr>
<td>Fluid</td>
<td>27</td>
<td>3</td>
</tr>
<tr>
<td>Symptoms</td>
<td>23</td>
<td>2</td>
</tr>
<tr>
<td>Orange Pigment</td>
<td>30</td>
<td>3</td>
</tr>
<tr>
<td>Margin &lt;3mm to disc</td>
<td>13</td>
<td>2</td>
</tr>
<tr>
<td>Ultrasonographic Hollowness</td>
<td>25</td>
<td>3</td>
</tr>
<tr>
<td>Halo Absence</td>
<td>7</td>
<td>6</td>
</tr>
</tbody>
</table>

To Find Small Ocular Melanoma Using Helpful Hints

<table>
<thead>
<tr>
<th>Thickness</th>
<th>Fluid</th>
<th>Symptoms</th>
<th>Orange</th>
<th>Margin to disc</th>
<th>Ultrasound</th>
<th>Halo</th>
</tr>
</thead>
<tbody>
<tr>
<td>U/S</td>
<td>O/S</td>
<td>P/P</td>
<td>S/S</td>
<td>O/S</td>
<td>U/S</td>
<td>M/S</td>
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<th>Symptoms</th>
<th>Halo</th>
<th>U/S</th>
<th>Hollowness</th>
<th>Disc distance</th>
</tr>
</thead>
</table>

USC Roski Eye Institute
Kern Medicine of USC
Which nevi grow?

- 27x greater risk ratio for 5 factors vs. 0 factors
- Growth not guarantee of malignancy
- Drusen are a sign of chronicity (favorable)
- Lifetime risk <1%

Shields - Chance of Growth at 5 years

Risk Factors

- Shields Combination of clinical factors
- If zero risk factors: 4% chance of growth/5 years
- If one risk factor: 36% chance of growth/5 years
- If 2 risk factors: >45% chance of growth/5 years
- If all risk factors: >56% chance of growth/5 years

- 27x greater risk ratio for 5 factors vs 0 factors
- Growth not guarantee of malignancy
- Drusen are a sign of chronicity (favorable)
Rumor #3: Everything that’s pigmented and elevated must be a melanoma or a high risk choroidal nevus
Diagnosis: PEHCR
(peripheral exudative hemorrhagic chorioretinopathy)

- Large choroidal and subretinal heme
- Elderly, caucasian patients
- Associated with drusen and blood thinner use
- Usually no trauma
- Often temporal
- Lumpy bumpy and cystic spaces on Bscan
Case

• 55 yo F with known breast cancer presents with sudden loss of vision

Diagnosis: Choroidal Metastases

• most common choroidal tumor in adults
• Women=Breast, lung, unknown
• Men=Lung, Unknown, GI
• Lung often precedes the systemic diagnosis
• Breast rarely does
• Poorly circumscribed, amelanotic, associated with subretinal fluid
• Can be bilateral and/or multifocal (20%)
Rumor #4: Everything that’s amelanotic must then be a met
Amelanoic choroidal nevus

Amelanotic melanoma

CHPRE and nevus in the other eye!
case

• 65 yo F with known breast cancer presents for routine evaluation
Breast cancer and primary Amelanotic choroidal melanoma

• Occam’s razor does not always apply
• Patients can have two primary cancers

Rumor #5: Rules exist for a reason (but they don’t always apply in Ocular Oncology…)}
Case

- 65 yo M with a history of a nevus with painless vision loss

One year ago

now
Rumor #6: Everything that grows is cancer
Diagnosis: BDUMP  
(Bilateral diffuse uveal melanocytic proliferation)

- rare paraneoplastic ocular syndrome
- benign hyperplasia of uveal melanocytes
- The GROWTH is not CANCER
- Painless bilateral vision loss
- Diffuse pigment clumping and orange pigment
- Subretinal fluid is common
- May precede diagnosis of systemic carcinoma by 3-12 months

Case

- A 45 year old male presents with a red, painless eye
Diagnosis: Ocular Adnexal Lymphoma

- Low grade Non-Hodgkins B-cell Lymphoma
  - 80% Extranodal marginal Zone lymphoma/Mucosa associated lymphoma
- Often affects the orbit, lacrimal gland, lids and conjunctiva
- Associated with systemic disease in 30%
- Conjunctival involvement is most associated with systemic disease
- Presents as a thick, velvety salmon patch

Do not confuse with OSSN!

No touch with cryo
Rumor #7: Ocular adnexal lymphoma is always an external disease.
Diagnosis: Uveal Lymphoma

- Low-grade Non-Hodgkins B-cell Lymphoma (MALT)
- Often affects the choroid, iris and/or ciliary body
- Prolonged indolent course often misdiagnosed as birdshot, white dots syndrome or VKH
- Key finding: yellow-white choroidal infiltrates with associated cresenteric choroidal thickening and hypofluoresence of ICG

- It is NOT vitreoretinal lymphoma (worse prognosis by far)

- 60% of patients with uveal lymphoma have OAL overlap
  - 50% are bilateral
  - 30% have systemic involvement
- Don’t fall for the rumor - Dilate patients with a salmon patch
Case

- 56 yo M presented for evaluation of a ‘spot’, recently started timolol in the right eye only
Diagnosis: Iris Nevus

• Common iris tumor
• Concern for melanoma with thickness >1 mm (average is 2mm), distortion of iris stroma, correctopia, ectropion uveae, feather borders, angle involvement
• Risk for malignant conversion ~8%
• Risk for metastatic disease is low ~3%
• Other high risk features: ABCDE
  – young Age, Blood (hyphema), inferior Clock hour, Diffuse, Ectropion uveae
  – High pressure also a risk factor
Rumor #8: Iris nevi are no big deal

Case continues

• Treated for recalcitrant unilateral glaucoma
case

• 58 yo Male from Egypt is referred for evaluation of a conjunctival nevus
Diagnosis: Primary Acquired Melanosis

- Painless, flat brown spot
- Often misdiagnosed as freckle or nevus
- Benign
- PAM with atypia – precancerous lesion with ~15% risk of progression to conjunctival melanoma
- Conjunctival melanoma ~50% mortality at 3 years, worse with >2mm, ulceration, caruncular involvement

Rumor #9: Conjunctival ‘nevī’ are no big deal either
case

• 28 yo Hispanic Female presents for evaluation of decrease vision x 6 months, worse after becoming pregnant
Diagnosis: Pigmented IridoCiliary Body Mass

• Often a late diagnosis
• May cause sectoral cataract
• Look for a sentinel vessel (important clue!)
• Considered a worse prognostic feature for uveal melanoma because it is detected later

Rumor #10: Ciliary body tumors are bad, bad, bad
Ciliary body adenocarcinoma
Ciliary body melanoma
Ciliary body melanocytoma

Ciliary body lymphoma
Ciliary body adenoma
Ciliary body adenocarcinoma

Ciliary body melanoma
Ciliary body melanocytoma
Case continues: Biopsy...

Pathology: adenoma
I love rumors. I always find out amazing things about myself - I never knew.

Thank you!