Update on the Management of Diffuse Choroidal Hemangioma

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Disclosures

• None
Introduction

• Diffuse choroidal hemangioma is an uncommon and benign type of vascular hamartoma
• Oftentimes associated with Sturge-Weber syndrome
• Characterized by diffuse choroidal thickening with possible overlying non-rhegmatogenous retinal detachment
• Cystoid macula edema (CME) may also be present
Patients and Methods

• Retrospective review of consecutive series of 11 patients (15 eyes) with diffuse choroidal hemangioma

• All patients had clinical features compatible with Sturge-Weber syndrome

• Patients evaluated between 08-2018 and 01-2020, and followed for 6 to 12 months

• Diagnosis established via history, external examination, indirect ophthalmoscopy, echography, and SD-OCT
Clinical Features

• Mean age 19.3 years
• Eleven patients (4 females, 7 males)
• Secondary glaucoma in eight patients
• Four patients had bilateral diffuse choroidal hemangiomas
• Two patients (2 eyes) had total exudative retinal detachment (RD) to the back of their lens, while a total of seven patients (10 eyes) had varying degrees of exudative RD
• VA variable ranging from 20/20 to HM
Options

Observation

Focal thermal photocoagulation

Anti-VEGF therapy

Photodynamic therapy (PDT)

External beam radiotherapy (EBRT), plaque radiotherapy, charged-particle radiation

Oral propranolol
Treatment Employed

• Photodynamic therapy (PDT) employed in six patients (6 eyes)
• External beam radiotherapy (EBRT) was utilized in two patients (2 eyes) followed by PDT
• Two patients (2 eyes) had EBRT alone
### Representative Case

<table>
<thead>
<tr>
<th></th>
<th>OD</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>VA</td>
<td>20/20</td>
<td>6/200</td>
</tr>
<tr>
<td>IOP</td>
<td>16</td>
<td>19</td>
</tr>
<tr>
<td>CRx (8/19)</td>
<td>-0.75 +0.50 x 090</td>
<td>+4.00 sph</td>
</tr>
<tr>
<td>EOM</td>
<td>Full</td>
<td>Full</td>
</tr>
<tr>
<td>Pupils</td>
<td>4 to 3 mm</td>
<td>Peaked pupil</td>
</tr>
</tbody>
</table>

**Anterior Segment OS**

- Tube in AC, unobstructed, no tube-cornea touch
- Positive posterior synechiae
- Anterior capsular pigment
Widefield fundus photo OU
Spectral domain OCT OS
Treatment

**Opted to employ**

Photodynamic therapy (PDT)
Treatment Response

PDT parameters
Spot size: 5000 μm x 3
minimally overlapping spots
Duration: 83 sec

<table>
<thead>
<tr>
<th></th>
<th>OS (pre-PDT)</th>
<th>OS (1 month post-PDT)</th>
<th>OS (3 months post-PDT)</th>
</tr>
</thead>
<tbody>
<tr>
<td>VA</td>
<td>6/200</td>
<td>20/70</td>
<td>20/50</td>
</tr>
<tr>
<td>IOP</td>
<td>19</td>
<td>19</td>
<td>17</td>
</tr>
</tbody>
</table>
1 month Post-PDT B-scan OS
3 month Post-PDT B-scan OS
Treatment response on SD-OCT

Pre-treatment

1 month

3 months
Response to Treatment

- All hemangiomas responded to the PDT and/or EBRT
- Two patients required a second treatment 5-6 months following the initial therapy
- Post-treatment VA varied widely, though all patients showed improvement
- Echographic measurements of choroidal thickness in the macular region dropped on average from 4.3 mm to 2.9 mm
Diffuse Choroidal Hemangioma

Background

- Usually ipsilateral to angiomatous malformation of skin (though may be bilateral)
- More likely to develop secondary RD spontaneously or following glaucoma filtering surgery
- Observed in 50% of patients with Sturge-Weber syndrome

Case example from W. Mieler
Diffuse Choroidal Hemangioma
Treatment Options

1. Photodynamic therapy (PDT)
2. Anti-VEGF therapy
3. External beam radiotherapy
4. Proton beam or stereotactic radiotherapy
5. Plaque brachytherapy (Iodine-125, Ruthenium-106, Cobalt-60)
6. Oral propranolol
Diffuse Choroidal Hemangioma

PDT

- Standard dose of verteporfin (6 mg/m$^2$), followed by application of 689 nm red laser light to lesion

- Variation in number of spots, spot size (2,500-7,700 μm), duration

- Avoiding extensive overlapping PDT spots → theoretical risk of increased fibrosis in area of overlap

- Generally treat the nodular area of the hemangioma

- There can be slow reabsorption of fluid, and possible initial worsening of exudative RD following treatment
Summary

• Treatment is individualized on a case-by-case basis

• In the presence of an overlying non-rhegmatogenous retinal detachment, PDT is generally deployed

• The entire extent of the choroidal hemangioma generally does not need to be treated

• Visual and anatomic outcomes tend to be favorable (though quite variable)


Thank You