An Unusual Case of Optic Disc Drusen

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14 year-old female presented with a history of decrease in vision. She states that she has noticed blurry vision in the right eye for 2 months. She believes her vision is progressively getting worse. She has no visual complaints in her left eye. Her last visit was 5 years ago and she has been seeing a general ophthalmologist since.
EXAM

• VA: 20/200 with no improvement on PH OD; 20/20 OS
• IOP 16 OD 14 OS
• Pupils: Reactive OU, No APD noted.
• Conj: white and quiet OU
• Iris: Normal OU
• Cornea: Clear OU
• Lens: Clear OU
• Anterior Segment: Deep and quiet OU
• Vitreous: Clear OU
• Optic nerve: optic nerve drusen with a grayish membrane temporal to the optic nerve OD; Optic nerve drusen OS
• Macula: subretinal hemorrhage nasal, thickening, watermark extends temporal to fovea OD; Normal foveal light reflex, no subretinal hemorrhage, no subretinal fluid OS.
• Vessels: Normal OU
• Periphery: Retina attached OU
OCT

OCT Retina: Subretinal thickening nasally with mild intra-retinal fluid. A small amount of subretinal fluid present OD. Unremarkable OS.

OCT Nerve: elevation of the optic nerve and hyper-reflective deposits OU.
**FA**

**OD:** Hyperfluorescence temporal to the optic nerve and an area of blockage nasal to the fovea is present. As the study progresses there is an increase in hyperfluorescence temporal to the optic nerve consistent with leakage.

**OS:** Staining of the optic nerve but no leakage in the macula.
B-Scan

Drusen present OU.
Fundus photography
Fundus Photography

OD: Subretinal thickening and fibrosis through the nasal macula with a subretinal hemorrhage nasal to the fovea.
Assessment and Plan

1. Choroidal neovascularization OD
2. Optic nerve drusen OU

Recommend anti-VEGF therapy OD for the choroidal neovascularization.
## Treatment Course

<table>
<thead>
<tr>
<th>Visit date</th>
<th>Vision OD</th>
<th>Treatment given</th>
<th>Visit Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>4/10/15</td>
<td>20/200 PH: NI</td>
<td>Initial treatment OD</td>
<td>As noted in previous slides.</td>
</tr>
<tr>
<td>5/12/15</td>
<td>20/200 PH 20/160</td>
<td>treatment scheduled for 5/28/15</td>
<td>Subretinal Heme resolved, subretinal thickening improve. Slight increase in subretinal fluid. Recommend IVA.</td>
</tr>
<tr>
<td>5/28/15</td>
<td>20/200 PH: NI</td>
<td>2nd anti-vegf treatment OD</td>
<td>Same as above</td>
</tr>
<tr>
<td>6/26/15</td>
<td>20/200 PH: 20/160</td>
<td>Pt denies further treatment.</td>
<td>Definite Improvement from initial visit. Still some subretinal fluid overlying the area of subretinal fibrosis. It is unclear if it will ever resolve. Recommended additional IVA to complete loading dose.</td>
</tr>
</tbody>
</table>
Fundus Photography: Post tx
Goldman VF OD

A hint of constriction nasally with slight enlargement of the blind spot.
Before & After

Before treatment

After treatment
Before & After

Before treatment

After treatment
Other case reports

Choroidal neovascularization secondary to optic nerve drusen is rare in children and literature suggests that further longitudinal studies are warranted in regards to treatment.

- Gregory Evans, et al. reports a case of a 11 year-old male that presented 20/100 OD and underwent 3 monthly anti-vegf treatments. VA improved to 20/20. ¹
- Alkin, et al. reports a 13 year-old female that presented with 20/80 OD vision. 1 month after 1 anti-vegf treatment VA improved to 20/25 and remained stable for 9 months. Knape report a case of bilateral choroidal neovascularization secondary to optic nerve head drusen. Initial VA was 20/200 OD and 20/70 OS. After the 1st IVA treatment VA improved to 20/60 OD and 20/70 OS. ²
- Delas, et al. 12 year-old female that presented with CF at 20 cm OD VA. She underwent two anti-vegf injections and VA improved to 20/60 and remained stable at 1 year. ³
Optic disc drusen

- Prevalence: 0.34% (clinically) 2% (on autopsy)
- No sex predilection
- Often bilateral (75-86%)
- Can be sporadic or AD
- Theories - impaired ganglion cell axonal transport, probably from small scleral canal or mechanical obstruction.
- Associated with retinitis pigmentosa and pseudoxanthoma elasticum
- In children optic disc drusen are generally buried and then become more visible throughout the years
- It is a cause of pseudopapilledema
- 8.6% of patients with optic disc drusen have transient visual obscurations
- 75-87% have a nerve fiber bundle defect.
- VF can show an enlarged blind spot, arcuate scotoma, or a sectoral scotoma.
Optic disc drusen

• Can cause serious ocular conditions in the adult population:\(^4\)
  • Central retinal vein occlusion
  • Central retinal artery occlusion
  • Peripapillary choroidal neovascularization
  • Ischemic optic neuritis
  • Retinal or optic nerve hemorrhage
• Choroidal neovascularization is a very rare complication in children from optic disc drusen. The proposed mechanism of the neovascularization is due to a compressive effect of the drusen on surrounding blood vessels leading to a compromised vascular integrity, vascular congestion, or ischemia.\(^4\)
  • A study in children with optic disc drusen showed delayed peripapillary choroidal filling in almost 50% of the patients.\(^5\)
Widely varying treatment options of choroidal neovascularization in children from optic disc drusen are found within the literature:

- Focal laser photocoagulation
- Photodynamic therapy with verteporfin
- Surgical removal of the membrane
- Recently Anti-Vegf therapy has been used off-label.
- A combination of focal laser and anti-VEGF has been reported by Knape et al.
- Observation - spontaneous involution has also been reported

There are thus far no studies comparing observation with the above reported treatment modalities.
References


