Optic Nerve Pit Maculopathy

Optic Nerve Pit (ONP)

- Incidence is 1/11,000
- Typically Unilateral
- Affected disc larger
- Grey or white oval depression
- Temporal disc location

Etiology of ONP

- May be due to incomplete closure of superior end of embryonic fissure – not proven
- Probably in a spectrum of disorders with optic nerve cavity disorders such as colobomas and morning glory syndrome. 
Optic Nerve Pit Histology

- Herniation of dysplastic retina through defect in lamina cribrosa

- Optic nerve fibers pass through lamina cribrosa, a collagenous sieve-like structure

- Thin membrane (remnants of Cloquet’s canal) overlies ONP

- **Arachnoid** layer surrounds optic nerve as exits the sclera creating subarachnoid space

- Arachnoid layer is middle layer (between dura and pia)

- Subarachnoid space contains cerebrospinal fluid (CSF)

- Space is continuous with brain
Optic Nerve Pit (ONP)

- VA unaffected unless macular detachment
- Macular detachment occurs in 30-50% of ONP
- Usually appears during 3rd/4th decade
- Chronic, slowly progressive VA loss

Optic Nerve Pit

- Average ONP size = 0.3DD
- Peripapillary atrophy present with RPE defect
- NFL and ganglion cell layer loss may lead to paracentral arcuate VF defect
Optic Nerve Pit vs Coloboma

- ONP has distinct margins
- Absence of CNS malformations in ONP so neuro-imaging not required.
- Coloboma often AD inheritance.
- VA loss in ONP only if macular detachment
- VA severely affected with ON Coloboma

Congenital Anomalies of Optic Nerve

Without Systemic Association
- tilted optic disc
- optic disc drusen
- optic disc pit
- myelinated nerve fiber

With Systemic Association
- optic disc coloboma
- morning glory syndrome
- optic nerve hypoplasia
- Aicardi syndrome
- megalopapilla
- peripapillary staphyloma
- optic disc dysplasia

Source of Fluid in ONP Maculopathy: Theory 1

- Source of fluid = liquefied vitreous
- Fluid movement from vitreous into/under any retinal layer through ONP
- Fluid creates “schisis” like cavity in outer retina (any layer possible)
- Chronic cystic changes in outer retina may create hole resulting in macular detachment
Source of Fluid in ONP Maculopathy

**Theory 1**

- Schisis-like cavity can occur in any retinal layer
- Macular detachment is secondary to the schisis-like separation
- Detachment occurs as subretinal fluid accumulates in subretinal space
- Schisis-like cavity connects to ONP but not to the macular detachment

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Source of Fluid in ONP Maculopathy

**Theory 2**

- Fluid source is from vessels at the base of the ONP
  
  But, FA fails to show extension of dye into subretinal space.
Source of Fluid in ONP Maculopathy

**Theory 3**
Leakage of CSF through ONP into subretinal space
unlikely due to:
- Thin membrane still exists at ONP between subretinal space and subarachnoid space
- Large difference in hydrostatic pressure between CSF and IOP

ONP Maculopathy

**Pathogenesis**
- Vitreous syneresis occurs
- Vitreous traction on macula creates negative pressure gradient
- Fluid draws into ONP—outer retina layers
- Fluid enters through outer wall hole or “percolates” into subretinal space
ONP Surgery

- PPV, induction of PVD, gas tamponade
- +/- peeling of ILM
- +/- peripapillary laser
- +/- drainage of subretinal fluid

- 72 year old Female presents with slow progressive VA loss OU

VA = 20/60 OD
VA = 20/200 OS

- mRX
  - OD: -10.00 +1.50 x 177
  - OS: -13.75 +1.00 x 110

OD
- (+) PVD
- Large staphyloma
- Focal RPE atrophy
- Juxtafoveal greenish, subretinal plaque with SRF
• NO PVD
• Staphyloma, peripapillary atrophy
• Temporal optic nerve pit
• Focal RPE atrophy
• Turbid submacular fluid
ONP Maculopathy OS

Diagnosis

• OD:
  — Subretinal neovascularization (SRN) due to pathological myopia

• OS:
  — Is submacular fluid due to SRN (high Myopia) or OPTIC NERVE PIT MACULOPATHY (ONP) ??
Factors in favor of SRN were:
1) Age (late for ONP)
2) High Myopia with SRN OD

However, minimal, if any Leakage on FA would be in favor of ONP as source of SRF

Management

• OD:
  – Avastin x 3 with complete resolution of leakage and SRF
• OS:
  – Avastin x 3 with no response

Diagnosis OS: ONP Maculopathy

• Treatment:
  Left PPV, ILM/ERM stripping, drainage of SRF, endolaser, Fluid gas exchange
  – MP relieves all traction on ONP to obtain closure
  – Laser creates barrier to fluid flow from ONP into retinal layers
  – Gas keeps subretinal space dry for laser effect
  – Resorption of SRF depends on RPE function(expect delay due to age and poor RPE function)
4 weeks PO

8 weeks Post PPV
- VA = 20/70
- Significantly less SRF
- OCT flatter
8 weeks Post PPV
• VA = 20/70
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