(Case Report)
Long-term Course of Chronic Pars Planitis

Remzi Avcı, MD, Professor
Ayşegül Mavi, MD, FEO, FICO
Sami Yılmaz, MD
Retina Eye Hospital-BURSA/TURKEY
ravci@bursaretina.com / www.bursaretina.com

Disclosure Statement
We have no financial relationships with any commercial interest related to the content of this presentation
Medical History

19 years old female, presented with complaints of bilateral visual loss (OD>OS)

Past Medical History
9 years ago (at the age of 10); diagnosis of Behçet Disease
  Azathioprine, cyclosporine, methotrexate, infliximab and adalimumab
5 years ago : Amnesia → Neurological assessment
  Council → Diagnosis of Neuro-Behçet

Ocular Examination
BCVA 20/20 OU, anterior segments quiet OU, IOP 14mmHG (OD) 12 mmHG (OS)

Past Medical History - 5 years ago

Posterior segment: vitritis OU

Multiple sclerosis related intermediate uveitis & vasoproliferative tumor
Medical History

**Treatment (1st Clinic / 2013-2017)**

- Azathioprine and interferon with a principal diagnosis of MS
- Bilateral intravitreal dexamethasone implant (3 years ago)
- Cataract surgery performed on right eye
- Bilateral 4 consecutive sessions of cryotheraphy
  
*last application 1 year ago*

Case Report

**BCVA : OD/OS → HM / 0.16**

*Slit lamp: Cells grade OD/OS → ++/+*
Preoperative Treatment Procedure

Megadose steroid therapy (1g) methylprednisolone i.v.
1mg/kg/day methylprednisolone p.o. for 10 days
VA : 20/20

All Surgical Interventions

OD 1-PPV + resection of tm + cryo + silicone oil tamponade
2-Silicone oil extraction + resection of tm + endolaser + C3F8 tamponade
OS 3-PPV + cryo + endolaser
4-Phaco/IOL
OD + OS 5-Strabismus surgery
Postoperative last visit (09.11.2018)

14 months after the first operation

- Hirschberg test : Orthophoric
- Slit lamp : Pseudophakia, AC quiet OU
- IOP : OD-OS $\rightarrow$ 15/16 mmHg
- BCVA : OD-OS $\rightarrow$ 0.2 – 1.0
- Fundus : Retina was attached, quiet, bilaterally. No vitritis/subretinal exudation was observed

Conclusions

- Timing of surgical intervention
  - Before permanent destruction of macula and retina
- Perioperative care
  - Preoperative control of uveitis before performing surgery
    Megadose i.v. Steroid $\pm$ oral steroid $\pm$ intraop. triamcinolone?
- Gradual surgical approach
Case Report: Long-term Course of Chronic Pars Planitis

Remzi Avci, MD, Professor
Ayşegül Mavi, MD, FEO, FICO
Sami Yılmaz, MD
Retina Eye Hospital-BURSA/TURKEY
ravci@bursaretina.com / www.bursaretina.com

Vasoproliferative tumor (VPT) - Introduction

- First description: Shields et al. 1983
- Classification: Idiopathic (%74)
  Secondary (%26)
VPT - Classification

Secondary VPT’s

- Believed to be a reactive vascular response to a variety of ocular insults. 3-6
  - Retinitis pigmentosa (22%)
  - Pars planitis (21%)
  - Coats Disease (16%)
  - Previous retinal detachment surgery (12%)


VPT – Clinical findings

- Sub-intraretinal exudations (%82)
- Exudative retinal detachment (%48)
- Vitreous hemorrhage (%21)
- Macular edema (%18) 3-5

VPT – Treatment alternatives

- **Cryotherapy**: Largest basal tm dia 10 mm> , tm thickness 5 mm> 1,3,12
- **Intravitreal anti-VEGF/ triamcinolone injection** 12
- **PDT** 9-11
- **PPV, tumor excision and endolaser** 3
- **Transscleral excision**: In cases of diagnostic difficulty to exclude the possibility of choroidal amelanotic melanoma 2
- **Plaque RT**: For large tumors or that have been resistant to cryotherapy 1,12
- **Enucleation**: For blind, painful eyes with NVG 2,3


VPT - Histopathology

- **Reactive, vascular-gliotic tissue** 3
- **Reactive gliosis**: A physiological response of CNS to conditions such as trauma (RD surgery), ischemia, inflammation (pars planitis), degeneration (retinitis pigmentosa) aiming to localize the lesion. 6-8
- **Reactionary retinal angiogliosis**
Conclusions

- **Irvine F et al**
  Pathological assessment
  The excised retinal mass, the attached choroid and inner scleral flaps were fixed in buffered 2.5% glutaraldehyde and processed for paraffin histological examination.

Microscopic examination demonstrated that the tumor consisted of a large mass of spindle cells surrounding blood vessels of various sizes. The spindle cells were of uniform size and mitotic figures were absent; positive staining with phosphotungstic hematoxylin and glial fibrillary acid protein confirmed the glial nature of these cells. The major part of vascular component took the form of clusters of channels lined by plump endothelial cells within hyalinized connective tissue. Fibrous, lipid and serous exudates were present in the tissue adjacent to the blood vessels. Extensive areas of 'basement membrane deposit' (synonyms: basal linear deposit, basal lamina deposit) were present on the inner surface of Bruch’s membrane. Elsewhere Bruch’s membrane was lined with a fibroreticulomembrane due to proliferation and metaplastic change within the RPE. These apperances were consistent with a peripheral retinal vascular tumor.

The histological features of these cases have allowed a more specific interpretation of these peripheral vascular lesions. We suggested that the new term 'reactive angiolysis' would seem a more appropriate descriptive term for the clinical entity previously classified as vasoproliferative retina tumors.

- **Khawly et al.**
  Because the weight of evidence in the literature supports the hypothesis that this pattern of pathology is reactionary, it is highly likely that proliferation of RPE seen in both of the specimens is also reactionary, secondary to a stimulus of unknown nature.

---

References


VPT - Histopathology

- **Reactive, vascular-gliotic tissue**
- **Reactive gliosis**: A physiological response of CNS to conditions such as trauma (RD surgery), ischemia, inflammation (pars planitis), degeneration (retinitis pigmentosa) aiming to localize the lesion. 6-8
- **Reactionary retinal angiogliosis**
- Furthermore none of VPT’s include a markedly dilated or tortuous feeding artery / draining vein as with the typical von Hippel retinal capillary hemangiomas. 3

---

Introduction

Vasoproliferative tumor (VPT)

- First description: Shields et al. 1983¹
- Pathology: involves not only the retina but occasionally RPE and choroid
- Histopathology: Mix of vascular, glial proliferation²
- Classification: Idiopathic (%74)
  Secondary (%26)³
- Locations: Between equator and ora serrata, inferior/ inferotemporal
- Findings: Sub-intraretinal exudations (%82)
  - Exudative retinal detachment (%48)
  - Vitreous hemorrhage (%21)
  - Macular edema (%18)³⁻⁵


Case Report:
Long-term Course of Chronic Pars Planitis

Remzi Avci, MD, Professor; Ayşegül Mavi, MD, FEBO, FICO; Sami Yılmaz, MD
Retina Göz Hospital-BURSA/TURKEY
ravci@bursaretina.com / www.bursaretina.com