

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



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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 \implies Neurological assessment

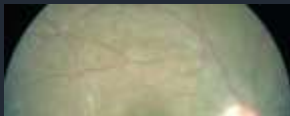
Council \implies Diagnosis of Neuro-Behçet

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



Courtesy of I T Tutkun

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

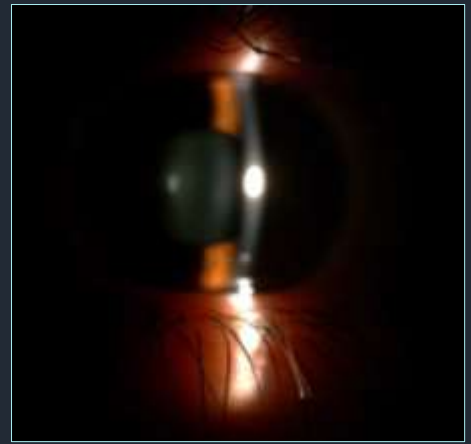
Bilateral 4 consecutive sessions of cryotherapy

(last application 1 year ago)

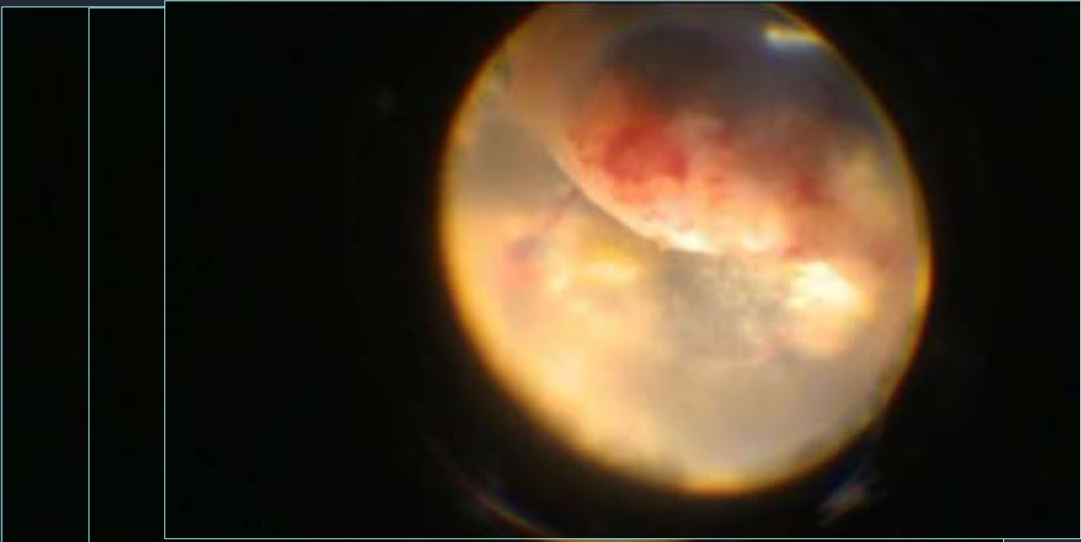
Case Report

BCVA : OD/OS \implies HM / 0.16

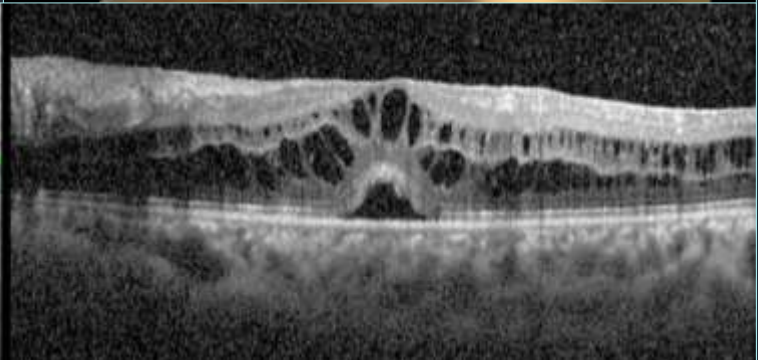
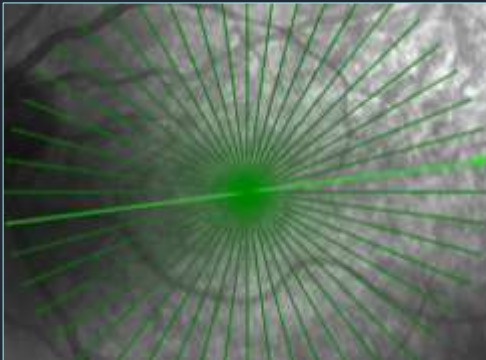
Slit lamp: Cells grade OD/OS \implies ++/+



Case - Fundus OD



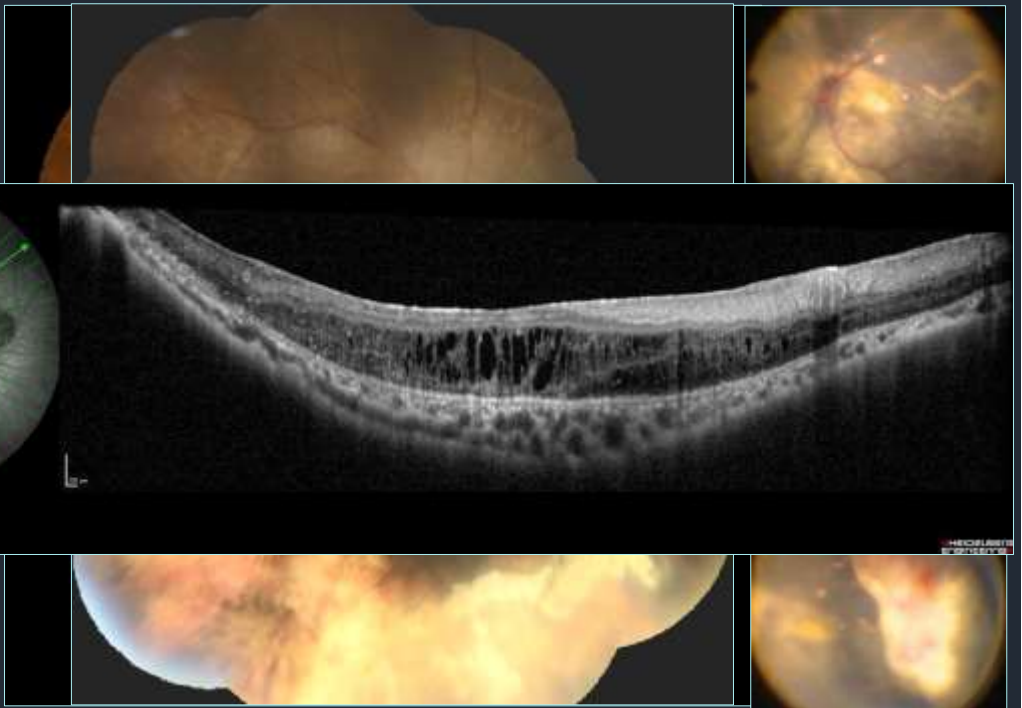
Case - Fundus OS

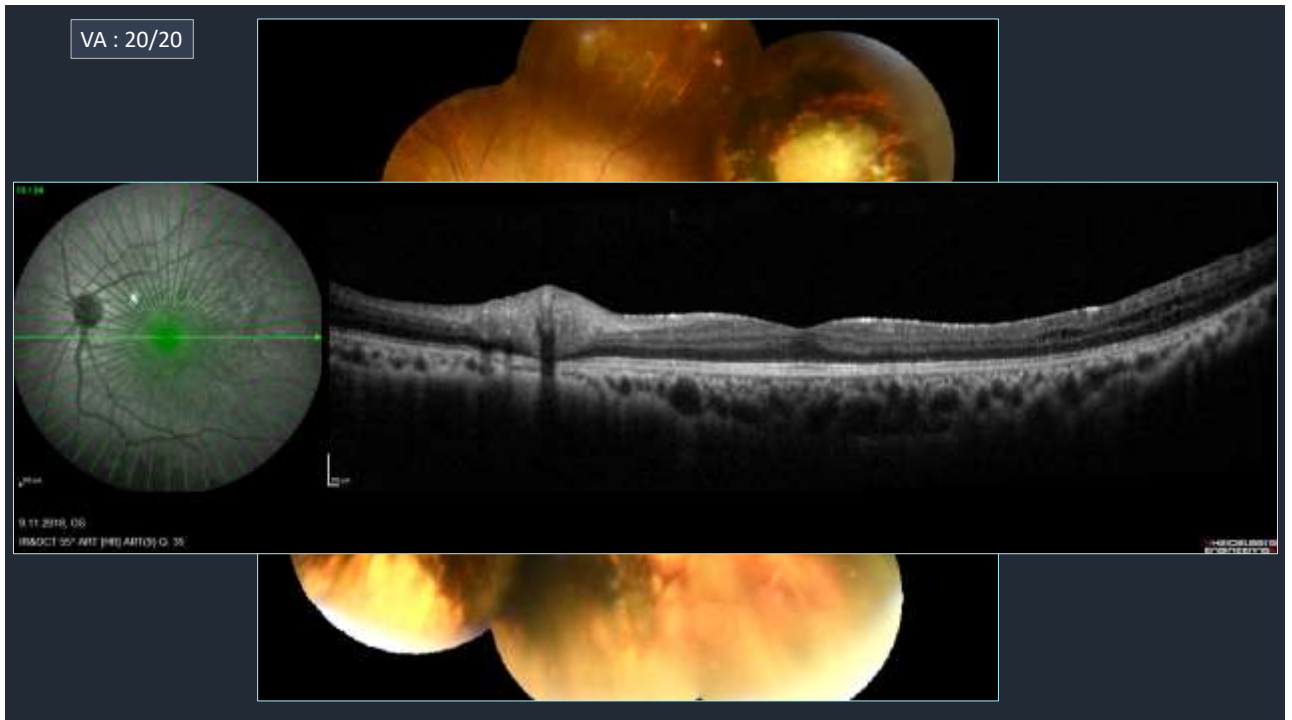


Preoperative Treatment Procedure

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

VA : 2/10





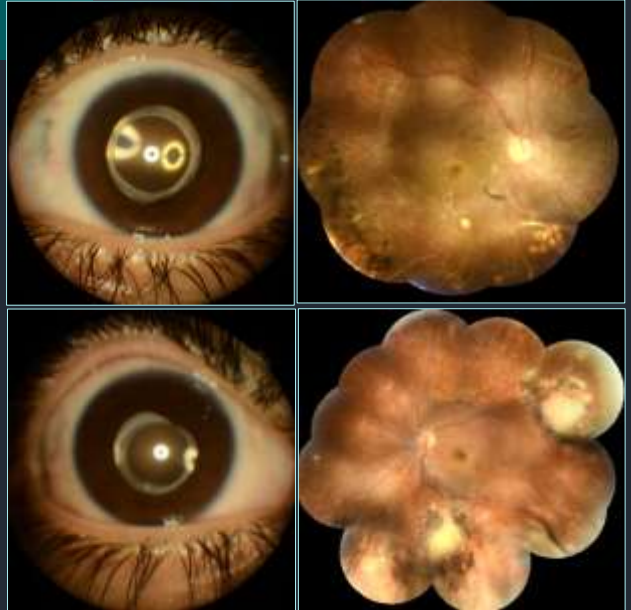
All Surgical Interventions

- | | |
|---------|--|
| OD | 1-PPV + resection of tm + cryo + silicone oil tamponade
2-Silicone oil extraction + resection of tm + endolaser + C ₃ F ₈ 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 \implies 15/16 mmHg
- BCVA : OD-OS \implies 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

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Vasoproliferative tumor (VPT) - Introduction

- First description : Shields et al. 1983¹
- Classification : Idiopathic (%74)
Secondary (%26)³

1. Shields JA, Decker WL, Sanborn GE, Augsburger JJ, Goldberg RE. Presumed acquired retinal hemangiomas. *Ophthalmology*. 1983;90(11):1292-1300.
2. Irvine F, O'Donnell N, Kemp E, Lee WR. Retinal vasoproliferative tumors: surgical management and histological findings. *Arch Ophthalmol*. 2000;118:563-569.
3. Shields CL, Shields JA, Barnett J, De Potter P. Vasoproliferative tumors of the ocular fundus: classification and clinical manifestations in 103 patients. *Arch Ophthalmol*. 1995;113(5):615-623.
4. Heimann H, Bornfeld N, Wj O, et al. Vasoproliferative tumours of the retina. *Br J Ophthalmol*. 2000;84(10):1182-1189.
5. Shields JA, Reichstein D, Mashayekhi A, Shields CL. Retinal vasoproliferative tumors in ocular conditions of childhood. *J AAPOS*. 2012;16(1):6-9.

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%)

3. Shields CL, Shields JA, Barrett J, De Potter P. Vasoproliferative tumors of the ocular fundus: classification and clinical manifestations in 103 patients. Arch Ophthalmol. 1995;113(5):615-623. 4. Heimann H, Bornfeld N, Vij O, et al. Vasoproliferative tumours of the retina. Br J Ophthalmol. 2000;84(10):1162-1169. 5. Shields JA, Reichstein D, Mashayekhi A, Shields CL. Retinal vasoproliferative tumors in ocular conditions of childhood. J AAPOS. 2012;16(1):6-9. 6. Shields CL, Kalki S, Al-Dannash S et al. Retinal Vasoproliferative Tumors. Comparative Clinical Features of Primary vs Secondary Tumors in 334 Cases. JAMA Ophthalmol. 2013;131(3):328-334.

VPT – Clinical findings

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

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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 ^{3,12}
- *Enucleation*: For blind, painful eyes with NVG ^{2,3}

¹ Shields JA, Decker WL, Sanborn GE, Augsburger JJ, Goldberg RE. Presumed acquired retinal hemangiomas. *Ophthalmology*. 1983;90(11):1292-1300., ² Irvine F, O'Donnell N, Kemp E, Lee WR. Retinal vasoproliferative tumors: surgical management and histological findings. *Arch Ophthalmol*. 2000;118:563-569., ³ Shields CL, Shields JA, Barrett J, De Potter P. Vasoproliferative tumors of the ocular fundus: classification and clinical manifestations in 103 patients. *Arch Ophthalmol*. 1995;113(5):615-623, ⁹ Blas MA, Scupola A, Tiberti AC, Sasso P, Balestrazzi E. Photodynamic therapy for vasoproliferative retinal tumors. *Retina*. 2006;26(4):404-409., ¹⁰ Chan RP, Lai TY. Photodynamic therapy with verteporfin for vasoproliferative tumour of the retina. *Acta Ophthalmol*. 2010;88(6):711-712., ¹¹ Barbesetto IA, Smith RT. Vasoproliferative tumor of the retina treated with PDT. *Retina*. 2003;23(4):565-567., ¹² Bianciotto C, Shields CL. Retinal vasoproliferative tumor with associated cystoid macular edema treated with cryotherapy and intravitreal triamcinolone. *Retina Today*. 2008:77-78

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

³ Shields CL, Shields JA, Barrett J, De Potter P. Vasoproliferative tumors of the ocular fundus: classification and clinical manifestations in 103 patients. *Arch Ophthalmol*. 1995;113(5):615-623, ⁶ Shields CL, Kaliki S, Al-Danmash S et al. Retinal Vasoproliferative Tumors. Comparative Clinical Features of Primary vs Secondary Tumors in 334 Cases. *JAMA Ophthalmol*. 2013;131(3):328-334., ⁷ Pekny M, Nilsson M. Astrocyte activation and reactive gliosis. *Glia* 2005; 50: 427-434., ⁸ Pekny M, Wilhelmsson U, Pekna M: The dual role of astrocyte activation and reactive gliosis. *Neurosci Lett* 2014; 565: 30-38.

Conclusions

- **Irvine F et al²**

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. Fibrinous, 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 by a fibrotic membrane due to proliferation and metaplastic change within the RPE. These appearances were consistent with a peripheral retinal vascular tumor.⁷

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

- **Khawly et al¹³**

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.

2. Irvine F, O'Donnell N, Kemp E, Lee WR. Retinal vasoproliferative tumors: surgical management and histological findings. Arch Ophthalmol. 2000;118:563-569. 13. Khawly JA, Matthews JD, Macherer R. Appearance and rapid growth of retinal tumor (reactive astrocytic hyperplasia) Graefes Arch Clin Exp Ophthalmol. 1999 Jan;237(1):78-81.

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- **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 angiogenesis
- 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

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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)
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3. Shields CL, Shields JA, Barrett J, De Potter P. Vasoproliferative tumors of the ocular fundus: classification and clinical manifestations in 103 patients. *Arch Ophthalmol*. 1995;113(5):615-623.

4. Heilmann H, Borfield N, Vij O, et al. Vasoproliferative tumours of the retina. *Br J Ophthalmol*. 2000;84(10):1162-1169.

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