OPTIC DISC PIT MACULOPATHY

Essay

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List of Abbreviations

Abbreviation	Meaning
AODP	Acquired optic disc pit
AF	Autofluorescence
BCVA	Best corrected visual acuity
C2F6	Perfluoroethane
C3F8	Perfluoropropane
CF	Counting fingers
CNS	Central nervous system
CSF	Cerebrospinal fluid
EDI-OCT	Enhanced depth imaging optical coherence tomography
FA	Fluorescein angiography
FAF	Fundus autofluorescence
FFA	Fundus fluorescein angiography
Hh	Hedgehog
ICGA	Indocyanine green angiography
ICP	Intracranial pressure
ILM	Internal limiting membrane
ILMP	Internal limiting membrane peeling
IR	Infrared
IS/OS	Inner segment / outer segment
logMAR	logarithm of minimum angle of resolution
MLF	Multilayered fluid
ORD	Outer retinal dehiscence
ORL	Outer retinal layer
PPV	Pars plana vitrectomy

PVD	Posterior vitreous detachment
RPE	Retinal pigment epithelium
SF6	sulfur hexafluoride
SRF	Subretinal fluid
VA	Visual acuity

Introduction

Optic disc pit (ODP) is a congenital optic disc abnormality that is first described by Wiethe in 1882. Optic disc pit usually appears as a round or oval, gray, white, or yellowish depression in the optic disc. It can be localized to any part of the optic disc, but has been reported most often in the temporal aspect **(Nicholson et al., 2011)**. Optic disc pit is considered a rare clinical entity, with an estimated prevalence of 1 in 11,000 people in general population. No sex or racial predilection has been observed. Optic disc pits have generally been thought to be sporadic, though several reports exist of autosomal dominant inheritance. They are considered to be bilateral in 10–15% of cases **(Hsu and Layton, 2015)**.

Clinically, Optic disc pit may remain stationary throughout life as a congenital abnormality, although about 60% of eyes with optic pits still have visual field deficits on perimetry including arcuate scotomas and enlarged blind spot **(Christoforidis et al., 2012)**. However, they most often become symptomatic and present with visual deterioration when they are complicated by maculopathy with serous macular detachment and/or macular retinoschisis, which

occurs in about 25%–75% of patients with optic pits, most commonly in their third or forth decade **(Wong et al., 2014)**.

The pathogenesis of optic disc pit maculopathy (ODPM) and the source of the retinal fluid remains controversial. A proposed mechanism by which maculopathy develops in association with optic disc pits is that fluid emanating from the optic nerve pit extends into various layers of the retina, including the subinternal limiting membrane space, ganglion cell layer, inner nuclear layer, outer nuclear layer, or subretinal space, with the outer nuclear layer most commonly affected **(Imamura et al., 2010)**. Many possible sources of this fluid have been proposed including; fluid from the vitreous cavity, cerebrospinal fluid originating from the subarachnoid space surrounding the optic nerve, fluid from leaky blood vessels at the base of the pit and fluid from the orbital space surrounding the dura. **(Georgalas et al., 2011)**.

The diagnosis of ODPM is based mainly on fundus examination and optical coherence tomography (OCT) imaging. Fundus examination demonstrates an ODP that is usually located temporally with a coexistent macular elevation. OCT is the most helpful diagnostic tool for ODPM, it has also provided great contribution to the investigation of

the mechanism by which optic disc pit-associated maculopathy occurs. Other diagnostic procedures include fluorescein angiography, fundus autofluorescence and visual fields **(Amador-Patarroyo et al., 2015)**.

Different treatment strategies for optic disc pit maculopathy have been attempted with varying success. Studies have shown that conservative management is associated with a poor outcome. As such, surgical approaches have become the preferred treatment modality for many practitioners **(Shah et al., 2014)**. Argon laser photocoagulation between the detached retina and the optic nerve has been used to seal and minimize the communication from the pit to the subretinal space, but it is often unsuccessful and repeated treatments are needed. Intravitreal gas injection alone can induce pneumatic displacement of the outer layer detachment; however, this effect may be temporary. Vitrectomy, with various adjunct procedures, including internal limiting membrane peel, laser photocoagulation and gas tamponade has been shown to improve vision in several cases **(Ooto et al., 2014)**.