

Optical coherence tomography in optic pit maculopathy

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Abstract

Purpose To report the optical coherence tomography (OCT) findings in patients with optic pit maculopathy.

Methods Twelve eyes of 12 patients with optic pit maculopathy were evaluated. Cross-sectional OCT images were correlated with findings from slit-lamp biomicroscopy and fluorescein angiography. The presence of retinoschisis and serous macular detachment was evaluated and the height of the serous detachment also measured.

Results Visual acuities varied between 20/400 and 20/63. The height of the serous macular detachment at the fovea was between 323 and 548 μm . OCT findings showed that ten (83.3%) patients had both retinoschisis and serous macular detachment, one (8.3%) patient had only retinoschisis and one (8.3%) patient had only serous macular detachment. None of the patients had macular hole or vitreoretinal traction.

Conclusion These findings support the concept of a bilaminar structure that contains retinoschisis and serous macular detachment. Our data also showed that in some patients, the sole component of maculopathy was serous macular detachment or retinoschisis.

Keywords Optic pit maculopathy · Optical coherence tomography · Retinoschisis · Serous macular detachment

Introduction

Optic pit is a congenital optic disc anomaly defined by Wiethe in 1882 [1]. This anomaly is usually found on the temporal part of the optic disc, and unless it causes maculopathy, most patients have good visual acuity and no discomfort. The prevalence of optic pit is 1/11,000; the prevalence of maculopathy caused by optic pit has been reported to vary between 25 and 75% [2, 3]. The mechanism of progression is still not fully understood, but recent studies have shown that it consists of internal retinal schisis and retinal detachment. This double-layered maculopathy is considered to result from a closure anomaly on the top corner of the embryonic fissure [4].

Optical coherence tomography (OCT) is an important imaging technique for the evaluation of optic pit maculopathy which, if left untreated, is known to result in significant reduction of vision [5–7]. To date, various treatment methods have been tried. In the

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present study, the OCT results of 12 eyes of 12 patients with an identified reduction of vision due to optic pit maculopathy were evaluated.

Methods

Twelve eyes of 12 patients with identified reduction of vision due to optic pit maculopathy were included in the study. The patients were 27–47 years of age (mean: 34.5 years), five (41.6%) of whom were men and seven (58.3%) of whom were women. The left eye was involved in seven (58.3%) cases and the right eye in five (41.6%). No abnormality was observed in the other eyes of the patients. The time between perceived reduction of vision and the visit to our clinic varied between 2 days and 4 months. None of the patients had any systemic disease or was on medication, and patient recollections did not indicate any family history of optic pit maculopathy. The visual acuity of the patients was measured with the EDTRS chart and, in addition to systemic eye examination, monochromatic and coloured photographs were taken with a standard fundus camera, and fluorescein angiograms were obtained with the Heidelberg Scanning Laser Ophthalmoscope (Heidelberg Engineering, Heidelberg, Germany). OCT images were obtained with a Zeiss OCT 3 (OCT model 3000; Carl Zeiss Ophthalmic System, Humphrey Division, Dublin, Calif.). In addition to assessing intraretinal changes in the tomographic cross-sections, we also assessed the presence of serous macular detachment. In cases with serous macular detachment, the height of the detachment at the fovea was measured on the OCT images by calculating the perpendicular distance between the lower limit of the retina and the retinal pigment epithelium in the detachment area.

Results

Patients enrolled in the study had normal anterior segment examination, pupillary reactions, colour vision and intraocular pressure. Refraction errors in the eyes with optic pit were as follows: -2.00 D in two (16.6%) eyes, -3.00 D in one (8.3%) eye, $+3.50$ D in one (8.3%) eye and emetropy in eight (66.6%) eyes. Fundus examination of the patients

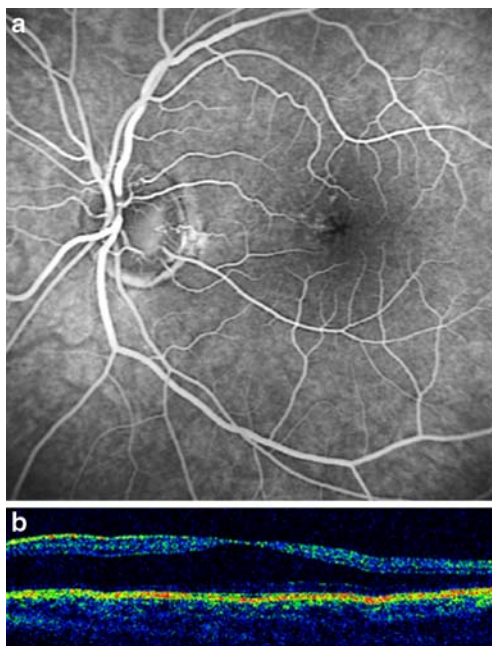
showed optic pit temporal to the optic disc. In addition, all patients had maculopathy due to optic pit. Fundus examination with a 78 D lens revealed serous elevation of the macula. An early hypofluorescein image of the optic pit showed dyeing at the late phase of fluorescein angiography. In patients whose OCT images showed serous macular detachment, early angiogram indicated hypofluorescence and late angiogram revealed oval or round hyperfluorescein lesions in the macula. In patients with an accompanying retinoschisis, irregular hyperfluorescein lesions were identified.

In 11 (91.6%) patients, OCT revealed the presence of a wide retinoschisis gap, observed as extensive hyporeflective areas in the retina. Multiple small cystic gaps were observed in the retina near the surface. In addition, 11 (91.6%) patients had serous macular detachment that caused convex subretinal hyporeflectance without compromising reflection in the retinal pigment epithelium underneath. OCT indicated only retinoschisis in case 1 and only serous macular detachment in case 2, whereas in the other cases, serous macular detachment was present with retinoschisis. The height of the detachment at the fovea was 203–548 μm in patients with serous macular detachment. Patients with retinoschisis had multiple vertical or oblique hyperreflective thin septa in the schisis cavity that extended from the retinal surface towards the retinal base. Retinoschisis was not limited to the fovea but extended towards the neighbouring tissues. Serous macular detachment did not extend towards the optic disc in any of the patients, and the images showed no macular hole or vitreomacular traction.

Visual acuity of the patients varied from 20/400 to 20/63. Case 1, whose OCT showed retinoschisis alone, had the highest visual acuity, while case 2, with the longest lapse of time between perceived reduction of vision and visiting the clinic and whose OCT showed serous macular detachment alone, had the lowest visual acuity. Since the number of patients enrolled in the study was not sufficient, statistical analysis of the height of serous macular detachment in relation to visual acuity was not possible. Table 1 presents the clinical findings, visual acuity measurements and OCT results. Fluorescein angiography and OCT images of case 1, case 2 and case 9 are shown in Figs. 1–3, respectively.

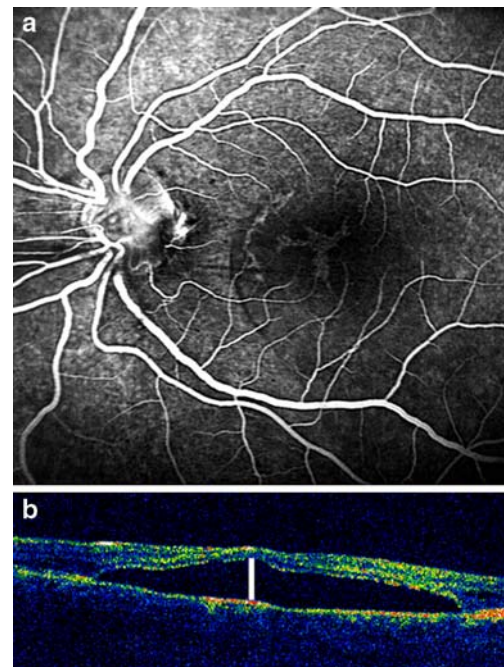
Table 1 Clinical findings, visual acuity measurements and optical coherence tomography (OCT) results of patients with optic pit maculopathy

Case no	Age	Sex	Duration of symptoms	Visual acuity	Retinoschisis	Serous macular detachment	Height of detachment (μm)
1	28	F	15 days	20/63	+	–	(–)
2	47	F	4 months	20/400	–	+	368
3	27	F	3 months	20/200	+	+	416
4	30	M	10 days	20/200	+	+	548
5	34	F	2 days	20/100	+	+	323
6	32	M	10 days	20/200	+	+	405
7	40	F	1 month	20/200	+	+	203
8	32	M	2 days	20/100	+	+	332
9	36	F	3 days	20/200	+	+	314
10	45	M	2 days	20/100	+	+	206
11	29	M	7 days	20/100	+	+	311
12	34	F	4 days	20/100	+	+	276

**Fig. 1** Fluorescein angiography (a) and horizontal optical coherence tomography (OCT) (b) images of case 1. The visual acuity was 20/63. The OCT image shows retinoschisis. There is no serous macular detachment

Discussion

As a congenital optic disc anomaly, optic pit is a rare clinical condition with a benign prognosis unless the macula is involved. Most of the cases are diagnosed

**Fig. 2** Fluorescein angiography (a) and horizontal OCT (b) images of case 2. The visual acuity was 20/400. The OCT image shows serous macular detachment. The height of the detachment at the fovea was measured on the OCT images by calculating the perpendicular distance between the lower limit of the retina and the retinal pigment epithelium in the detachment area. Note that there is no retinoschisis

during routine ocular examination. However, optic pit patients may develop maculopathy, which leads to significant reduction of vision [3]. Because optic pit

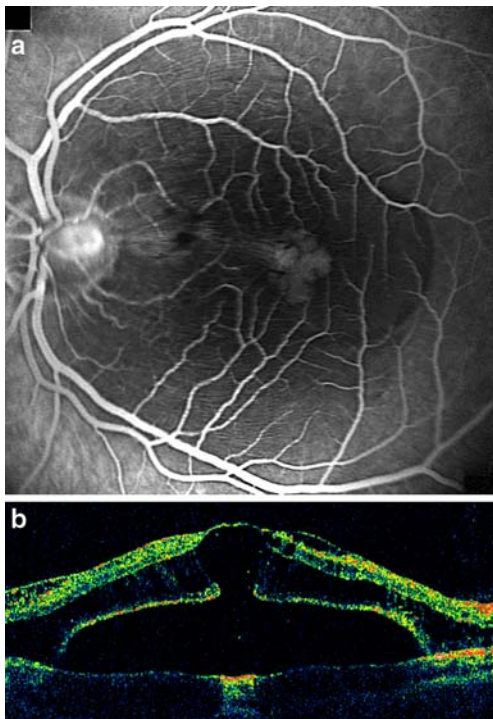


Fig. 3 Fluorescein angiography (a) and horizontal OCT (b) images of case 9. The visual acuity was 20/200. The OCT image shows retinoschisis and serous macular detachment. The height of the detachment at the fovea was measured on the OCT image by calculating the perpendicular distance between the lower limit of the retina and the retinal pigment epithelium in the detachment area. The height of the detachment at the fovea is 314 μm

is a congenital anomaly with a silent clinical prognosis until the onset of maculopathy, which leads to significant reduction of vision, it is important to understand the factors that cause maculopathy. Various theories have been developed about the onset of maculopathy. In terms of the pathogenesis of the detachment of the macula, Gass supported the view that the fluid is derived from the cerebrospinal fluid [8]. Bonnet and Joko claimed that vitreoretinal traction played an important role in maculopathy [9, 10]. Other investigators have considered that a microhole developing on the membrane covering the pit may push the vitreous fluid below the retina [11, 12]. The proponents of this theory claim that vitreoretinal traction may play a role in the development of this microhole and that hyaloid dissection during vitrectomy may improve the clinical condition [11, 12]. Conversely, Lincoff et al. claimed that as a component of maculopathy, retinoschisis develops

before the onset of macular detachment and that the fluid in the schisis cavity travels to the subretinal gap and leads to serous detachment within the macula [4]. This group supported their proposal on the basis that the serous detachment is limited to the macula and the detachment has no connection with the optic disc [4]. Based on fluorescein angiography findings, Gass suggested that prognosis would be worse in cases with serous macular detachment and that patients with long-term untreated serous macular detachment were prone to a significant loss of vision [13].

Different methods have been tried to treat optic pit maculopathy, a clinical condition with an unclear origin and a poor prognosis if untreated. The results obtained with laser photocoagulation are far from satisfactory [14]. Gas injection with or without vitrectomy has also been tried, with a success rate that is slightly higher than that obtained with laser photocoagulation; however, the recurrence of maculopathy has been observed, especially in the eyes with no vitrectomy procedure [15, 16]. Posterior scleral buckling is yet another method, but it is difficult and has varying success rates [17]. Some investigators have reported better results by combining vitrectomy, posterior hyaloid dissection, laser photocoagulation and gas injection [18].

The main reason why a widely accepted treatment method with documented improvement in visual acuity has not been reported to date is probably due to our lack of knowledge on the onset mechanism of maculopathy. OCT is an important imaging technique that enables the clinician to understand the clinical outlook of optic pit maculopathy as well as other types of maculopathy, thanks to the in-vivo assessment possibility it offers [19]. OCT helps us to follow the relation between the retinal surface and both vitreous fluid accumulation and schisis-like changes in the retina, and subretinal fluid accumulation and changes in the retinal pigment epithelium. Furthermore, as an easy technique with a high replication rate, OCT facilitates the follow-up of maculopathy patients.

In our study, the detachment of intraretinal structures and a wide retinoschisis gap in 11 of the 12 patients were revealed as extensive hyporeflective areas identified by OCT. Isolated serous macular detachment was observed in one patient, and isolated retinoschisis was observed in another patient; none of the patients showed vitreomacular traction or macular hole. In a similar study published by Rutledge et al.,

serous macular detachment was observed in only one of four eyes with optic pit maculopathy [7]. These investigators reported retinoschisis in all of the eyes and a lamellar hole in one eye. The detachment area in the eye with serous macular detachment was limited to the macula and was not directly associated with optic pit. A similar inference was made in another study conducted by Lincoff et al. [6]. In our series also, all serous macular detachments were limited to the macula. Interestingly, in case 2, OCT did not reveal any retinoschisis but only serous macular detachment. Case 2 was the patient who had the longest interval between a perceived reduction of vision and visiting the clinic, at which time the visual acuity was limited to 20/400. The variation observed in our series and in other studies should be emphasized [6, 7, 19]. It is known that optic pit maculopathy regresses spontaneously [20], and studies have shown that both retinoschisis and serous macular detachment regress with time. This is a slow process and takes many years in some patients. The alterations in the retina and retinal pigment epithelium caused by maculopathy usually result in reduced visual acuity. The time between the perceived reduction of vision and visiting the clinic varied between 2 days and 4 months in our patients. Furthermore, if we consider that maculopathy could have started even before the patient perceived a reduction in vision, it becomes apparent that the study group was not a homogeneous group with respect to the progression of maculopathy. It should also be noted that as an unknown pathology with unknown triggering factors, optic pit maculopathy may lead to different lesions in different eyes. In addition, in the present study, the OCT findings of only one examination were assessed, the changes in the lesions were not followed up and the lesion characteristics in relation to visual acuity in such a limited series could not be statistically analyzed. Despite these factors, our study showed that OCT is an extremely important imaging technique for the evaluation of optic pit maculopathy.

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