Original Article

Spectral domain optical coherence tomography morphology in optic disc pit associated maculopathy

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Purpose: Our purpose was to study the clinical manifestation and course of optic pit maculopathy using Spectral Domain Optical Coherence Tomography (SD- OCT) images. **Materials and Methods:** We used SD-OCT to examine 20 eyes of 19 patients with a macular detachment in combination with an optic. **Results:** We observed five different fovea appearances in regard to fluid localization. In five eyes, we recorded changes in the fluid distribution with SD-OCT. In 17/20 eyes, we noted a communication between the perineural and subretinal and/or intraretinal space at the margin of the optic disc. **Conclusion:** 3-dimensional SD-OCT (3D-SDOCT) scans revealed a three-fold connection, between subretinal and intraretinal space, perineural space, and the vitreous cavity. Therefore, we suppose that intraretinal or subretinal fluid in optic pit maculopathy may have both a vitreous and cerebrospinal origin. A membrane, covering the optic nerve was noted in 14 cases. Even if it seems intact in some B-scans, it is not complete in others several micrometers apart. Additionally, we observed fluid accumulation below the margin of the optic disc and hyperreflective porous tissue in the optic disc excavation. Those findings do not influence the course of maculopathy.

Key words: Maculopathy, optic pit, spectral domain optical coherence tomography

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Optic pit, described for the first time by Wiethe in 1882,[1] is a congenital colobomatous defect of the optic nerve head. At the beginning of the twentieth century, Reis estimated that the prevalence of this abnormality was as rare as 1 in 11,000 patients.[2] Optic pits occur mostly at the temporal side of the optic disc and are bilateral in 10-15% of cases.[3] In only about 20% of cases, visual prognosis without treatment is good (visual acuity over 0.75), mostly in patients with no abnormalities in the macular area.[4] In other patients, the optic pits were accompanied by central serous retinopathy, macular edema, macular cyst, macular hole, macular hemorrhage or progressing nerve fiber layer defects.[4-6] Patients with optic pits do not usually have any visual complaints until changes in the macular region develop, generally at the age of 21-30 years.[5] Pars plana vitrectomy seems the method of choice, but surgical details are still being discussed.[7-10]

Spectral Domain Optical Coherence Tomography (SD-OCT) visualizes the retinal structure by measuring the interferometric signal detected as a function of optical frequencies. It enables a 10100 times faster imaging speed than standard Optical Coherence Tomography (OCT).

Because of the low prevalence of the disease, controversies exist concerning the origin of fluid and mechanism of optic disc pit associated maculopathy.

The aim of this study is to present the clinical appearance of optic pit maculopathy in SD-OCT and its changes over time. Additionally, to present several macular and optic disc features using SD-OCT.

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Materials and Methods

This is an observational study, of consecutive patients with biomicroscopic and fluorescein angiography appearance of maculopathy combined with optic pit. We obtained Institutional Review Board approval and the study was conducted in accordance with the Declaration of Helsinki.

All patients were examined with SD-OCT (Spectralis, Heidelberg Engineering, Germany or SOCT Copernicus HR, Optopol, Zawiercie, Poland). In Copernicus HR, we performed 100 consecutive B-scans on a 7 × 7 mm area. The Spectralis device makes 19 B-scans on an area of 4.5 × 6 mm. We always took two scans, one presenting the macula and the other presenting the optic disc. In SD-OCT the central transverse scan was included to subgroup analysis each time. We then analyzed each individual scan in the raster pattern to determine additional features.

We recorded any changes in SD-OCT appearance and/or visual acuity that occurred during the observation period.

Results

19 patients (20 eyes) with maculopathy associated with optic disc pit were included into the study. The mean age of the 5 women and 14 men was 31 years. Mean follow-up was 24 months.

We sorted patients into five groups according to the localization of the fluid. Group 1 consisted of eyes with intraretinal fluid in outer retinal layers (three eyes), Group 2; eyes with intraretinal fluid in outer retina layers and subretinal fluid (three eyes), Group 3; eyes with intraretinal fluid in outer and inner retinal layers (five eyes), Group 4; eyes with an outer lamellar macular hole coexisting with fluid in inner and outer retinal layers (three eyes), and Group 5; eyes with elevation of all retinal layers in the macula (six eyes) [Fig. 1]. In five eyes intraretinal fluid migrated during the observation

period. The SD-OCT appearance of one eye from Group 1 (fluid in outer retinal layers) fluid migrated to the inner retinal layers and to the subretinal space [Fig. 2 a-c] and in four eyes from Group 3 the SD-OCT macular appearance changed to Group 4 [Fig. 2 d and e].

3-dimensional SD-OCT (3D-SDOCT) imaging of the optic nerve head revealed the presence of a membrane in the bottom of the optic disc in 14out of 20 cases. In only one case was the membrane complete, with no defects in all 3-D scans of that eye. In the remaining 13 eyes, although the 3-D imaging mode showed the membrane to be complete in some scans, in others it did not [Fig. 3].

In SD-OCTposterior hyaloid was visible in four cases with partial attachment to the optic nerve.

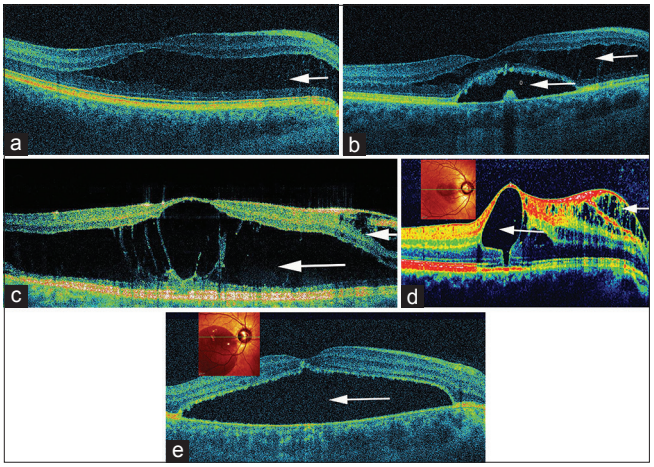

Figure 1: Five appearances of fluid accumulation in maculopathy associated with optic pit. (a) Fluid in outer retinal layers, (b) Fluid in outer retinal layers and subretinal space, (c) Fluid in outer and inner retinal layers, (d) Outer lamellar macular hole and fluid accumulated in inner and outer retina. On the left side infrared image of the fundus is presented, (e) Subretinal fluid without intraretinal fluid accumulation. On the left side infrared image of the fundus is presented

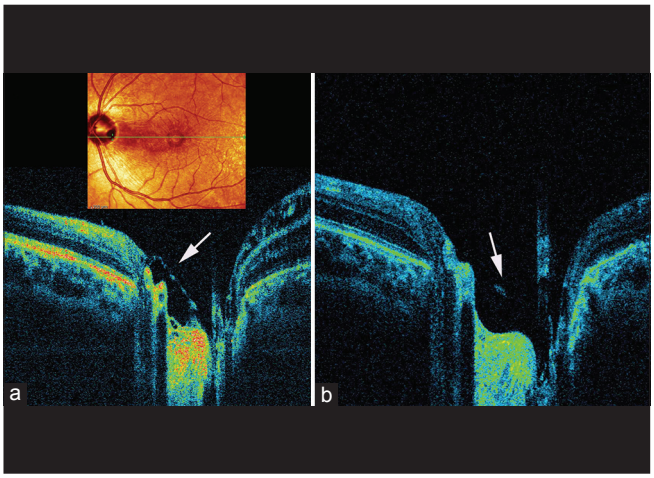

Figure 3: B-scans of the same patient 0.14 mm apart. The arrow on a indicates a membrane spanning over the optic disc. The arrow on b indicates a hypererreflective area, possibly a part of the membrane visible on a. On the left side above the SD-OCT images infrared image of the fundus is presented

A communication between the perineural and subretinal and/or intraretinal space at the margin of the optic disc existed in 17 out of 20 cases [Fig. 4].

SD-OCT revealed several abnormalities of the optic disc, which have previously only been described histopathologically. First, the presence of hyporeflective spaces below the optic disc was observed in nine cases [Fig. 5]. Whether these spaces represent an additional allocation of the perineural fluid, which

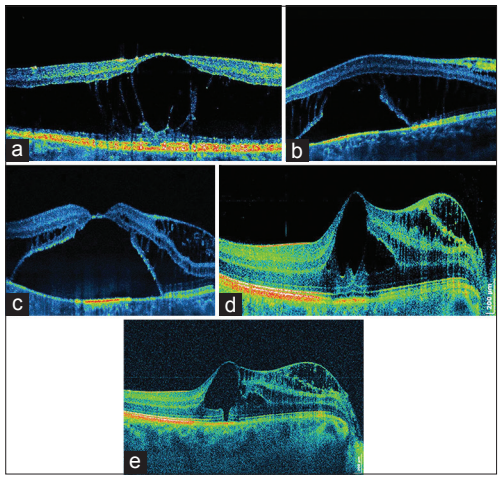

Figure 2: Changes in the fovea appearance and fluid distribution over time. (a-c) Maculopathy associated with optic pit in one patient visualized with spectral OCT. On scan A elevation of inner retinal layers, retino schisis and stria between inner and outer retinal layers can be seen. Visual acuity was 0.5 at that time. Scan B taken one year later. The stria between inner and outer retinal layers tend to shorten and outer retinal layers are detached from retinal pigment epithelium and pulled towards already detached inner retinal layers. Visual acuity improvement to 0.8 was noted at the time the scan was taken. On Scan C, further movement of the outer retinal layers towards the inner retinal layers can be seen. (d and e) Maculopathy associated with an optic pit in another patient. On scan D, intraretinal fluid is visible in inner and outer retinal layers. Scan E: Four months later an outer lamellar macular hole developed

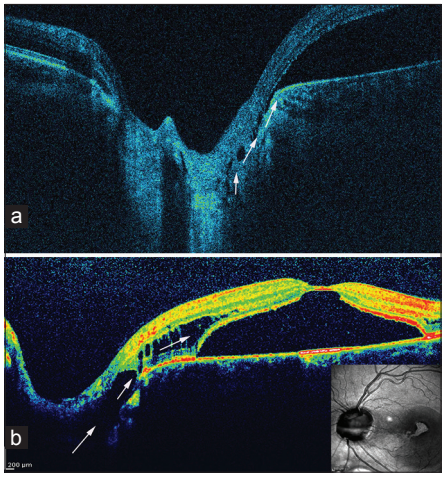

Figure 4: A communication between the perineural and: a, subretinalspace in a patient with maculopathy combined with optic pit. b, intraretinal space in a patient with maculopathy combined with optic pit. On the bottom right side infrared image of the fundus is presented. Arrows indicate the suspected flow of fluid

did not pass to the intraretinal and/or subretinal space, or that they represent also fluid accumulated below the Elschling membrane is open to discussion. Secondly, in eight cases hyperreflective tissue was observed at the bottom of the optic nerve [Fig. 6]. This tissue may represent either glial cells or condensed vitreous.

Discussion

This paper describes the morphology of the optic nerve and macula in optic pits visualized with 3D-SDOCT. As maculopathy associated with optic pit is a rare event, many factors remain unknown.

Origin of fluid

The origin of the subretinal and intraretinal fluid is a primaryinterest. Avitreous origin was confirmed by histopathologic studies showing that alcian blue staining revealed mucopolysacharydes within the pit.[5] Further endorsement of the argument for a vitreous origin of the fluid is offered by the good results following pars plana vitrectomy (PPV), which enables the relieving of tractions.[11] At the 2007 American Academy of Ophthalmology (AAO) Film Festival Anderson *et al*. presented "Optic Nerve Pit Bubbles", a movie showing gas bubbles coming out from the optic nerve in optic pit maculopathy previously treated with PPV with gas. Johnson and Johnson presented silicone oil moved to subretinal space.[12] All of the above confirm an interconnection between the vitreous and intraretinal space.

Cerebrospinal origin of the fluid that enters the subretinal and intraretinal space was also confirmed.[3,13,14] Chang observed that contrast dye can pass from subarachnoid space to subretinal fluid in humans with morning glory syndrome, which is generally considered as another manifestation of optic pit.[14] Kuhn *et al.* described a case of MRI-documented intracranial silicone oil migration in an eye with optic pit.[15]

Our SD-OCT images confirm that intraretinal fluid may be both, cerebrospinal and vitreous in origin. We think that there may be a three-fold connection between the vitreous, perineural space and subretinal and intraretinal space. It may be that in individual cases, the paths of fluid differ in size and trajectory. Vitrectomy, while relieving tractions may prevent the migration of vitreous fluid through the optic nerve into the subretinalor intraretinal space. Laser burns create a scar between the outer retinal tissue and retinal pigment epithelium and this may prevent migration of fluid into the outer retinal space, but does not influence fluid migration into inner retinal layers. As multiple possible fluid pathways exist, vitrectomy and/or laser coagulation may be successful in some cases, and yet may fail in others. Thus, an ideal procedure would combine the relieving of traction and prevention of fluid migration into the subretinal as well as into inner and outer retinal layers.

Appearance and origin of maculopathy

We observed that fluid may be observed in the outer retinal layers, both in the outer retinal layers and subretinal, in outer and inner retinal layers, in a form of an outer lamellar macular hole or only subretinal [Fig. 1]. We additionally presented evolution of the fluid distribution with time [Fig. 3]. The second much discussed issue is whether maculopathy is a primary process, or is associated with posterior hyaloid detachment. Some authors believe that macular detachment occurs only secondary to posterior hyaloid detachment, consisting either of detachment of the outer retinal layers from the retinal pigment epithelium without communication to the optic disc, or macular schisis with accumulation of fluid in the outer plexiform layer.[16-21] Other authors state that the posterior hyaloid remains attached in the majority of eyes, so macular detachment may be regarded as a primary process coexisting with the optic pit.[11,22-24] We confirmed in this SD-OCT study that partial vitreous detachment was observed only in 4/20 eyes. The role of SD-OCT in the detection of posterior hyaloid detachment may be controversial. Also, our observations during vitrectomy for optic pit associated maculopathy show that one of most difficult maneuvers was the induction of posterior hyaloid detachment (unpublished data). A confirmation of the above thesis may be the fact, that we observed evolution of maculopathy in five cases without any signs of posterior hyaloid detachment. Additionally, after vitrectomy, when the posterior hyaloid is already removed, the macula was reportedto reattach in several cases, which shows that vitreoretinal tractions are of minor importance in subretinal

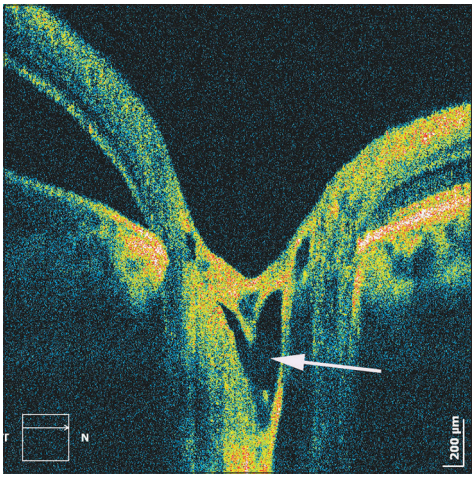

Figure 5: Optic disc appearance in a patient with optic pit associated maculopathy. The arrow indicates a hyporeflective area, possibly representing fluid accumulated below the optic nerve head

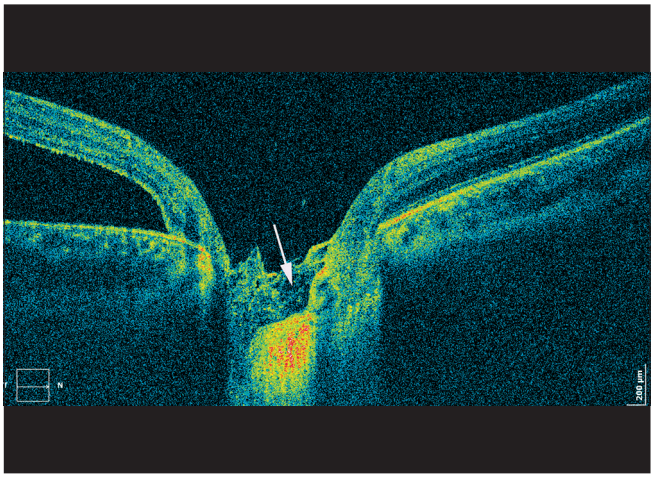

Figure 6: Optic disc appearance in a patient with optic pit associated maculopathy. The arrow indicates hyperreflective, porous tissue visible in the excavation of the optic disc

fluid accumulation. In view of these findings, we assume that macular detachment is primary to vitreous detachment. Vitreous may also detach when the macula flattens. Furthermore, we have never observed vitreous traction intraoperatively.

Morphology of the optic nerve

The third interesting aspect is the morphology of the optic disc itself. SD-OCT studies to-date have not paid much attention to this. We observed a connection between the perineural and sub-/or intraretinal space in 17 out of 20 cases [Fig. 4]. Earlier authors did observe such a connection, but not with such a high frequency, probably because of worse resolution of the OCT devices used in earlier studies.[25] In most cases in our group we observed a membrane in the bottom of the optic pit. In histopathologic studies a structure called the "inner limiting membrane of Elschnig" (a continuation of internal limiting membrane over the optic disc) was observed on top of the optic disc in optic pit maculopathy. This membrane was described to consist of rudimentary retinal tissue including aberrant nerve fibers and pigmented tissue resembling retinal pigment epithelium.[5] In an OCT based study, Doyle and coworkers identified a complete membrane traversing the optic disc cup in three of five eyes without maculopathy. The authors suggest that the membrane, consisting of neuroectodermal and astroglial tissue, may represent a barrier to the passage of fluid into or under the retina. As the membrane was absent or deficient in three eyes with maculopathy, they also suggested that the membrane may protect against the development of maculopathy.[26]

When we analyzed 3D-SDOCT images, we noticed that even if the membrane seems intact in some B-scans, it is not complete in others several micrometers apart. This might not have been noticed by Doyle and coworkers as they did not perform 3-dimensional reconstruction.[27]

In eight patients in our study we observed hyperreflective tissue in the excavation of the optic disc [Fig. 5]. This SD-OCT finding may be either condensed vitreous or glial tissue as explained by histopathological studies. Akiba observed a condensed vitreous strand (Cloquet's canal) that extended from the surface of the pit into the vitreous gel.[11] Cloquet's canal pulsates with eye movements forming multiple microforamina in the membrane covering the optic pit. The liquefied vitreous is believed to enter the subretinal space through the optic pit. Also, liquefied vitreous at the top of the optic pit was commonly reported.[3,11,27] Other studies claim that optic pits are filled with glial tissue.[4,5] Some authors hypothetized that this glial tissue may contribute to fovea detachment.[28,29]

In our study, SD-OCT revealed the presence of previously unreported hyporeflective areas at the bottom of the optic disc in nine cases [Fig. 5]. We suspect that these spaces represent an additional allocation of the perineural fluid, which did not pass to the intraretinal or subretinal space, although it is also possible that they represent fluid accumulated below the Elschlnig's membrane.

In conclusion, 3D-SDOCT scans revealed a three-fold connection, between subretinal and intraretinal space, perineural space, and the vitreous cavity. Therefore, we suppose that intraretinal or/subretinal fluid in optic pit maculopathy may have both a vitreous and cerebrospinal origin. It may enter the subretinal space at the optic disc cup near to the vessels outcome. Several other abnormalities of the optic disc are visible with SD-OCT, such as a membrane overlying the bottom of the optic disc (Elschlnig's membrane) which does not seem to have a role in visual prognosis. Additionally, we observed fluid accumulation below the margin of the optic disc and hyperreflective porous tissue in the optic disc excavation.

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