

Spectral domain optical coherence tomography imaging in optic disk pit associated with outer retinal dehiscence

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Abstract: A 37-year-old Bangladeshi male presented with an inferotemporal optic disk pit and serous macular detachment in the left eye. Imaging with spectral domain optical coherence tomography (OCT) revealed a multilayer macular schisis pattern with a small subfoveal outer retinal dehiscence. This case illustrates a rare phenotype of optic disk maculopathy with macular schisis and a small outer retinal layer dehiscence. Spectral domain OCT was a useful adjunct in delineating the retinal layers in optic disk pit maculopathy, and revealed a small area of outer retinal layer dehiscence that could only have been detected on high-resolution OCT.

Keywords: retinal imaging, maculopathy, macular schisis

Introduction

An optic disk pit is a congenital abnormality of the optic nerve head, first described in 1882 by Wiethe¹ as a gray, white–yellow, or black round depression in the optic disk.² Optic disk pits are extremely rare, with an incidence of one in 11,000.³ In 1927, Halbertsma⁴ described an association of optic disk pit with macular lesions and poor visual acuity, which is now known as optic disk pit maculopathy (ODP-M). ODP-M can develop in 25%–75% of optic disk pits and usually confers a poor visual prognosis, with 80% of eyes having a final visual acuity of 6/60 or worse, as a result of long-standing serous macular detachment, formation of macular holes, and atrophy of the underlying retinal pigment epithelium.⁵ The morphology of ODP-M has been well characterized by the use of optical coherence tomography (OCT).⁶ ODP-M has a typical bilaminar appearance of retinal schisis and neurosensory detachment, but, commonly, either one of the features exists in isolation. Outer retinal holes or dehiscence are uncommon findings in ODP-M, reported to occur in only 27% of cases⁶ imaged with time domain OCT (TD-OCT). Spectral domain OCT (SD-OCT) offers high-resolution imaging of the retinal layers, allowing a detailed analysis of the morphology of ODP-M, and may detect small areas of outer retinal dehiscences missed on TD-OCT imaging. We describe a case of ODP-M with a small area of outer retinal dehiscence that was revealed on SD-OCT.

Case report

A 37-year-old Bangladeshi male with no past medical history of note presented with a central blurring of vision in his left eye since 1 month prior to his visit to the ophthalmology clinic. His best-corrected visual acuity (BCVA) on the Snellen chart was 20/20 in the right eye and 20/400 in the affected left eye. The anterior segment

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examination was unremarkable, and the intraocular pressure on Goldmann applanation tonometry was 18 mmHg in both eyes. A dilated fundal examination of the left eye revealed neurosensory detachment of the macula with scattered subretinal deposits within the area of detachment. An optic disk pit was seen in the inferotemporal aspect of the optic disk, continuous with the area of retinal detachment (Figure 1). The optic disk and fundus of the right eye were normal. SD-OCT (Spectralis HRA-OCT; Heidelberg Engineering, Heidelberg, Germany) revealed outer nuclear layer macular schisis with neurosensory detachment and a small area of outer retinal dehiscence in the foveal region (Figure 2). The outer nuclear layer schisis cavity was communicating with the optic disk pit (Figure 3). Autofluorescence imaging showed isoautofluorescence in the macula with multiple hyperautofluorescent spots (Figure 4) corresponding to the lipofuscin deposits seen on fundal examination. Fundus fluorescein angiography showed temporal parapapillary hyperfluorescence due to leakage from the optic disk pit (Figure 5). A diagnosis of ODP-M with outer retinal dehiscence was made. Treatment options, including surgery, were discussed. The patient was not keen for surgical intervention due to cost issues, and eventually underwent focal argon laser photocoagulation to the temporal optic disk margin. Unfortunately, he did not return for his subsequent review.

Discussion

Our case illustrates several typical OCT and autofluorescence features of ODP-M. First, our patient had intraretinal fluid accumulation within the outer nuclear layer, which is the most common site of involvement in ODP-M.⁷ Second, there was coexisting neurosensory detachment. This dual morphology is another common finding in ODP-M.⁷ Third,

autofluorescence imaging revealed multiple hyperautofluorescent subretinal deposits, which are an almost universal feature of ODP-M.^{8,9} An uncommon finding in our case was that of a small area of subfoveal outer retinal dehiscence that was picked up on high-resolution SD-OCT.

The etiology of ODP-M remains controversial. It is postulated that the possible sources of the intraretinal/subretinal fluid could be the vitreous cavity,⁹ cerebrospinal fluid from the subarachnoid space,¹⁰ leaky blood vessels at the base of the optic disk pit,¹¹ or the orbital space surrounding the dura.¹² How or why this fluid tracks into the retina is unknown. Postel et al¹³ proposed that a rhegmatogenous mechanism might be the triggering event by which liquefied vitreous enters the optic disk pit via a small hole in the overlying diaphanous tissue.¹⁴ Lincoff et al¹⁵ suggested a sequence of events in the natural history of ODP-M, beginning with fluid accumulation in various retinal layers, with subsequent development of outer retinal dehiscence and movement of fluid through the dehiscence into the subretinal space.¹⁴

Contrary to Lincoff et al's findings,¹⁷ outer retinal dehiscence may not be a mandatory step for the occurrence of subretinal fluid accumulation. Most of the earlier studies describing features of ODP-M with TD-OCT do not report the presence of an outer retinal dehiscence,^{5,15–19} with few exceptions.^{6,20} A recent study by Imamura et al utilized OCT to characterize ODP-M in a consecutive series of 16 patients and found outer retinal dehiscence in only 27% of eyes and presence of subretinal fluid in some patients without outer retinal holes.⁶ However, a variety of OCT machines were used in the study, including TD-OCT and SD-OCT machines. TD-OCT may not have the requisite resolution for imaging small outer retinal holes, possibly contributing to an underestimation of their incidence.

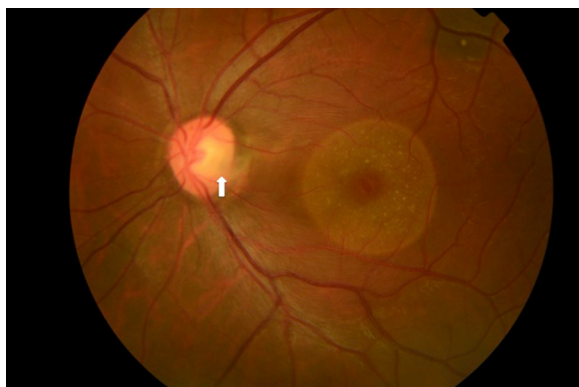


Figure 1 Fundus photograph of the left eye.

Note: The image shows macular neurosensory detachment with multiple scattered subretinal deposits and an inferotemporal optic disk pit (white arrow).

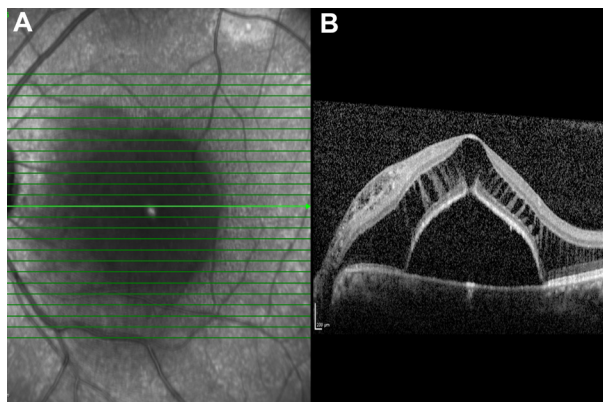


Figure 2 Spectral domain optical coherence tomography scan of the left macula showing outer nuclear layer schisis, a small area of outer retinal dehiscence, and the presence of subretinal fluid.

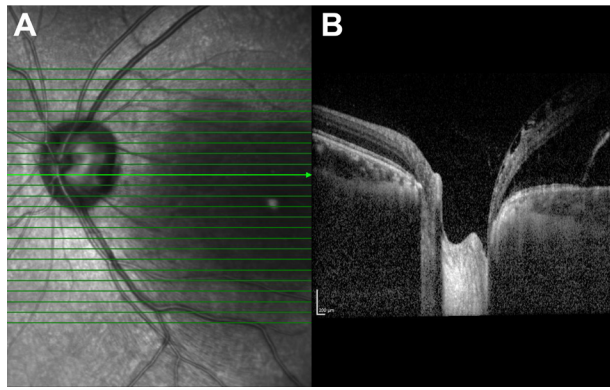


Figure 3 Spectral domain optical coherence tomography scan showing communication between the optic disk pit and the outer nuclear layer schisis cavity.

Twenty-five percent of ODP-M cases resolve spontaneously,² but the poor visual outcome of conservative management²¹ has prompted use of a more aggressive approach. The treatment options range from barrage laser photocoagulation to vitrectomy, with or without adjunctive procedures such as internal limiting membrane (ILM) peel and gas tamponade. Argon laser photocoagulation is applied to the disk margin in the area of the retinal detachment and is believed to cause full-thickness retinal scarring that impedes the flow of fluid, thus allowing the retinal pigment epithelial pump to reabsorb existing fluid.⁶ In most cases, reattachment of the retina occurred, but over a prolonged period of up to 1–2 years.^{22,23} The release of anterior–posterior traction with pars plana vitrectomy and tangential traction with ILM peel has shown promising anatomical and functional outcomes.^{7,14} Shukla et al performed vitrectomy with ILM peeling, barrage laser photocoagulation, and gas tamponade in a prospective series of seven patients with macular schisis and central outer retinal dehiscence.²⁴ Good



Figure 4 Fundal autofluorescence image of the left eye showing multiple hyperautofluorescent spots within the area of macular detachment.



Figure 5 Fundal fluorescence angiogram of the left eye showing parapapillary hyperfluorescence at the temporal optic disk margin.

visual outcomes were achieved, with a median preoperative BCVA of 20/60 improving to a median postoperative BCVA of 20/30. However, four out of seven patients developed full-thickness macular holes (FTMH) postoperatively. The authors attributed the high incidence of FTMH to the peeling of ILM over thinned-out retina. FTMH as a postoperative complication following surgery for ODP-M was similarly reported in other studies.^{13,21} In the absence of strong evidence for any particular treatment, it may be prudent to adopt a graded approach, starting with argon laser photocoagulation and progressing to surgery if the maculopathy fails to resolve. It is also unknown if morphological features, such as multilayered schisis, outer retinal dehiscence, or neurosensory detachment, may necessitate earlier or more aggressive treatment for better visual outcome. Our patient was offered surgical treatment but opted for a more conservative approach of laser photocoagulation first.

Conclusion

We present a rare phenotype of ODP-M with macular schisis and neurosensory detachment in the presence of a small area of outer retinal dehiscence. SD-OCT was a useful adjunct to diagnosis in ODP-M, and revealed a small area of outer retinal layer dehiscence and the extent of macular schisis. Further studies are needed to explore the significance and impact of structural features in ODP-M on the choice of treatment and visual prognosis.

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Disclosure

The authors report no conflicts of interest in this work.

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