Peripapillary Detachment in Pathologic Myopia

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Objective: To describe peripapillary detachment in pathologic myopia (PDPM), a newly recognized fundus lesion.

Design: Retrospective medical record review.

Methods: We evaluated a series of myopic eyes that had a yellow-orange elevation of the retina and retinal pigment epithelium at the inferior border of the myopic conus.

Results: Twenty eyes of 15 patients were identified during a 17-year period to have characteristic findings of PDPM. The mean age of the patients was 58 years. They were followed up for an average of 6 years. The mean spherical equivalent correction was –11.00 diopters (D) (range, –6.00 to –16.00 D). The mean axial length was 27.4 mm (range, 25.3-28.9 mm). In each case, ophthalmic coherence tomographic examination showed a localized detachment of the retinal pigment epithelium and retina corresponding to the PDPM lesion. During the follow-up period, the lesion remained stable in all cases except for 1. No apparent negative effect on visual function was noted.

Conclusions: Peripapillary detachment in pathologic myopia is an asymptomatic, yellow-orange peripapillary detachment of the retinal pigment epithelium and retina in pathologic myopia. Recognition of this lesion is important to distinguish it from other fundus pathologic conditions, such as tumors or choroidal neovascularization, which require further investigation and treatment.

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MYOPIA IS the most common ocular abnormality. Pathologic myopia is defined by a refractive error of –6.00 diopters (D) or greater and an axial length of the globe of more than 26 mm.[1] Progressive chorioretinal degeneration is often associated with pathologic myopia.[2]

Fundus changes in pathologic myopia are numerous and are related to the degree of myopia, axial length, and the presence of a posterior staphyloma. The expansion of the globe with scleral thinning leads to the formation of a posterior staphyloma and optic disc myopic conus.[3]

The clinical features of the myopic fundus have been well described. The purpose of this report is to describe a newly observed lesion at the inferior margin of the peripapillary conus in a series of patients with pathologic myopia. In 1984, we first noticed the presence of an elevated, well-circumscribed, dome-shaped, yellow-orange lesion inferior to the optic disc along the inferior margin of the myopic conus. This lesion appeared to be subretinal because the retinal vessels passed over it. When viewed stereoscopically, it appeared to be a small localized retinal elevation or thickening. We called this lesion "peripapillary detachment in pathologic myopia" (PDPM). We first reported the presence of a PDPM lesion in a smaller case series, which was presented at the Association for Research in Vision and Ophthalmology annual meeting in 1995.[4] For this study, the original patients as well as additional cases identified since were asked to return for further assessment, including optical coherence tomographic (OCT) examination of the area of PDPM. This report summarizes our experience during almost 2 decades with this peculiar fundus abnormality in pathologic myopia.

METHODS

During a 17-year period from January 1984 to December 2001, 15 patients with pathologic myopia and PDPM in one or both eyes were identified. Medical records from these patients were reviewed for the following data: age at initial examination, sex, race, number of eyes

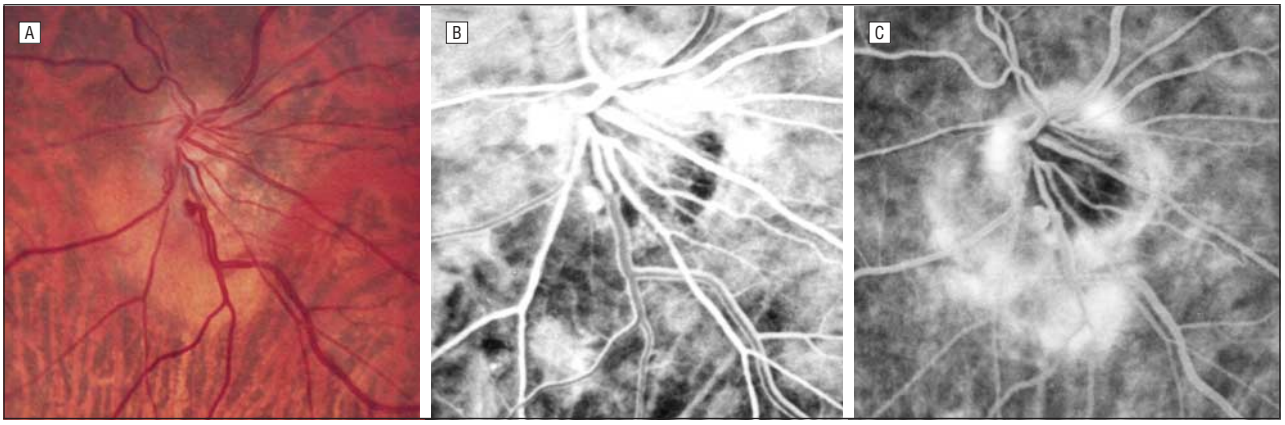

Figure 1. Case 1. A, Photograph of the left eye of a 73-year-old woman demonstrates the presence of an elevated, yellow-orange lesion (peripapillary detachment in pathologic myopia [PDPM]) at the edge of a myopic conus. There is an anomalous vasculature that appears to be an inferonasal vein emanating from the peripapillary retinal pigment epithelial lesion rather than from the disc. B, Early fluorescein angiogram of the same eye reveals hypofluorescence in the area of PDPM. This hypofluorescence was interpreted as blocked fluorescence because the underlying choroidal vessels cannot be seen through the lesion. C, Late-phase fluorescein angiogram demonstrates hyperfluorescence consistent with staining of the PDPM.

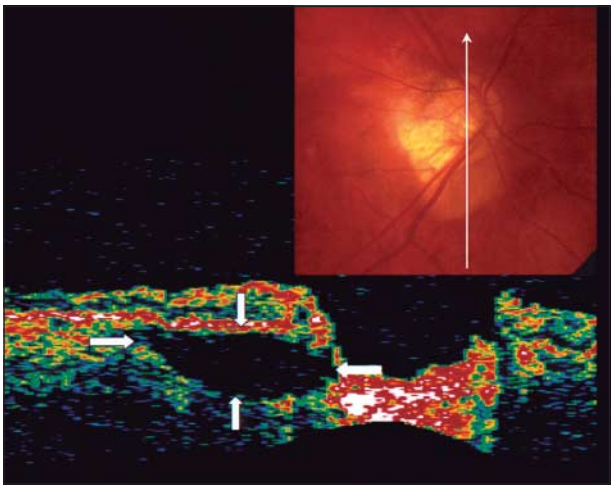

Figure 2. Case 2. Ophthalmic coherence tomographic image of the right eye of a 71-year-old man demonstrates the presence of a localized detachment that appears to involve both the retinal pigment epithelium and retina (thick white arrows).

involved, refractive error, best-corrected visual acuity, visual symptoms, and associated myopic fundus changes, such as tilted disc, posterior staphyloma, myopic conus, lacquer cracks, and Fuchs spot. A-scan axial length measurements and results of B-scan ultrasound examination, color fundus photography, visual field testing, and fluorescein angiographic examination, when present, were recorded. All of the patients were asked to return to the retinal research department of the Manhattan Eye, Ear, and Throat Hospital (New York, NY) for a follow-up visit. Each patient underwent a complete ophthalmologic examination, including best-corrected visual acuity measurement, intraocular pressure measurement, anterior segment biomicroscopy, and dilated fundus examination. Furthermore, color fundus photographs were obtained and each patient underwent fluorescein angiographic examination, A-scan axial length measurement, B-scan ultrasound evaluation of the posterior segment, and OCT examination of the peripapillary area. We performed OCT examinations with commercially available OCT equipment (Humphrey-Zeiss, Dublin, Calif) developed from the prototype described by Hee et al[5] and Puliafito et al.[6] For each patient, we performed scans at multiple angles overlying the lesion. The area of transition from the peripapillary detachment to the optic disc was studied with additional scans at varying angles. Microperimetric examination of the area of PDPM with a scanning laser ophthalmoscope was performed in 4 of 15 patients; another 4 patients underwent visual field testing (Humphrey 30-2). The following is a detailed presentation of several representative cases drawn from this series.

CASES

Case 1

A 73-year-old woman was examined for bilateral decreased visual acuity. She was pseudophakic in both eyes but she reported high axial myopia since high school and manifested typical features of the myopic fundus. She had a history of hypertension and hypothyroidism. Her best-corrected visual acuity was 20/25 OD and 20/20 OS. The right eye had a tilted disc with a peripapillary myopic conus. The left eye had a yellow inferonasal peripapillary elevated lesion at the inferior edge, clearly distinct from the patient's myopic conus (**Figure 1**A). The choroidal vessels could not be seen beneath the lesion. An anomalous vasculature was present, with an inferonasal vein emanating from the peripapillary RPE lesion rather than from the optic disc. The fluorescein angiographic examination showed initial hypofluorescence with late staining of the lesion (Figure 1B and C). The OCT showed a nonreflective area that appeared to be beneath both the RPE and retina corresponding to the area of PDPM. There were no changes in the PDPM at the 1-year follow-up. Visual acuity remained at 20/20 OS.

Case 2

A 71-year-old man was evaluated for background diabetic retinopathy. The mean spherical equivalent correction was –6.00 D OD and –2.00 D OS. Mild nuclear sclerosis was present bilaterally. In the right eye, there was a small, tilted disc, a temporal conus, and an adjacent inferior shallow yellow-orange elevation, consistent with PDPM. The left optic disc was normal. The OCT through the PDPM demonstrated the presence of a well-circumscribed area of elevation, which appeared to involve both the retina and RPE (**Figure 2**). An oblique scan through the PDPM and the myopic conus revealed what appeared to be a full-thickness defect in the retina-RPE layers (**Figure 3**). This patient was followed up for 1 year, and no change was noted in the PDPM in the right eye.

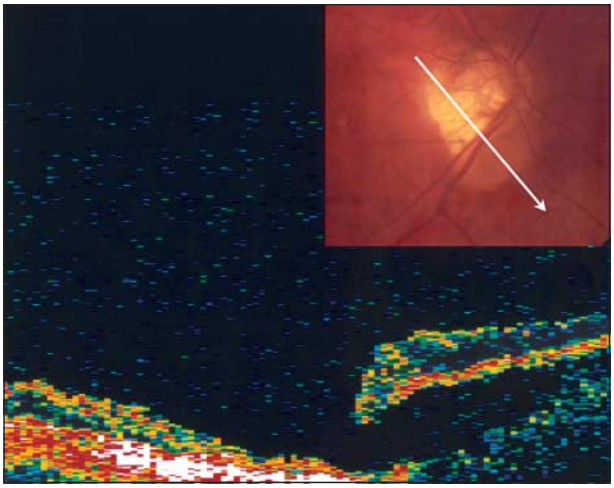

Figure 3. Case 2. Optical coherence tomographic image at an oblique angle reveals the presence of a full-thickness discontinuity in the retina–retinal pigment epithelial layers at the margin of the peripapillary detachment in pathologic myopia in the same patient.

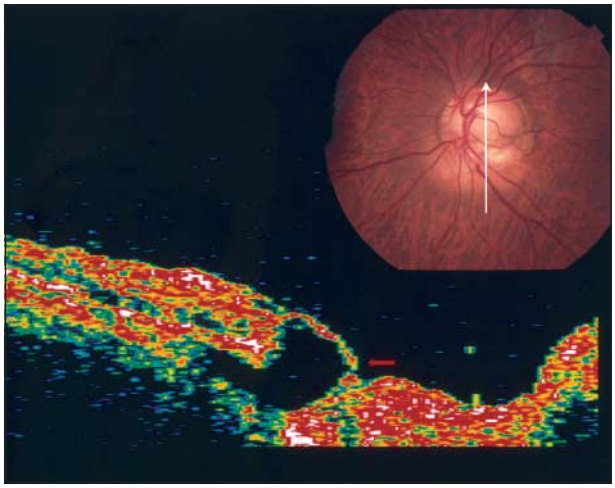

Figure 4. Case 3. Clinical photograph and optical coherence tomographic (OCT) scan of a 48-year-old man showing peripapillary detachment in pathologic myopia (PDPM) lesions at the inferior border of the patient's myopic conus. The lesion obscures the underlying choroidal vasculature. The OCT image reveals the presence of a pigment epithelial detachment corresponding to the PDPM. The inferior retinal veins appear to dip beneath the superior edge of the lesion. Note the thinning of the retina at the superior edge of the PDPM on the OCT scan (red arrow).

Case 3

A 60-year-old man was referred for a decline in vision in the right eye for 2 months. His best-corrected visual acuity was counting fingers OD and 20/20 OS. The mean spherical equivalent correction was –16.00 D OD, and –9.25 D OS. On fundus examination, there was an area of subfoveal choroidal neovascularization in the right eye. In both eyes, there was a yellow-orange retinal elevation inferior to the myopic conus. Optical coherence tomographic examination over the area of retina elevation confirmed the presence of a PDPM in both eyes. In the left eye, the OCT scan revealed the presence of retinal thinning, with almost a full-thickness defect at the edge of the PDPM (**Figure 4**). The patient was followed up for 18 months, and no change was detected in the PDPM lesions.

Case 4

A 76-year-old man was referred for evaluation of degenerative myopia and choroidal neovascularization. His best-corrected visual acuity was 20/400 OD and 20/30 OS, with a spherical equivalent correction of –10.00 D OU. In the right eye, there was a disciform macular scar but no PDPM. In the left eye, there was a small, tilted disc surrounded by a myopic conus and a yellow-orange, well-demarcated elevation inferior to the disc, consistent with a PDPM. A fluorescein angiogram of the left eye revealed initial hypofluorescence and late staining of the area of PDPM and subfoveal choroidal neovascularization (CNV). An OCT examination confirmed the presence of a detachment in which the neurosensory retina and the RPE appeared elevated (**Figure 5**). The patient was followed up for 4 years without any change in the PDPM. The subfoveal CNV in the left eye progressed to a disciform scar during 1 year. His visual acuity in the left eye decreased to 20/400. Fluorescein angiographic examination demonstrated late staining of the PDPM throughout the follow-up period (**Figure 6**).

Case 5

A 62-year-old man was referred for evaluation of pathologic myopia. Four years earlier, he had developed a nonischemic central retinal vein occlusion in the left eye. He was subsequently found to have carotid artery obstruction and underwent endarterectomy bilaterally. The visual acuity was 20/20 OD, and 20/60 OS. In the right eye, there was a tilted disc, myopic conus, and a yellow-orange, well-circumscribed elevation contiguous with the myopic conus. The lesion obscured the underlying choroidal vasculature. An OCT scan revealed the presence of a localized detachment (**Figure 7**). A 3-dimensional B-scan ultrasound confirmed the presence of a localized elevation just inferior to the optic nerve (**Figure 8**). The patient was followed up for 3 years with no changes noted in the PDPM in the right eye.

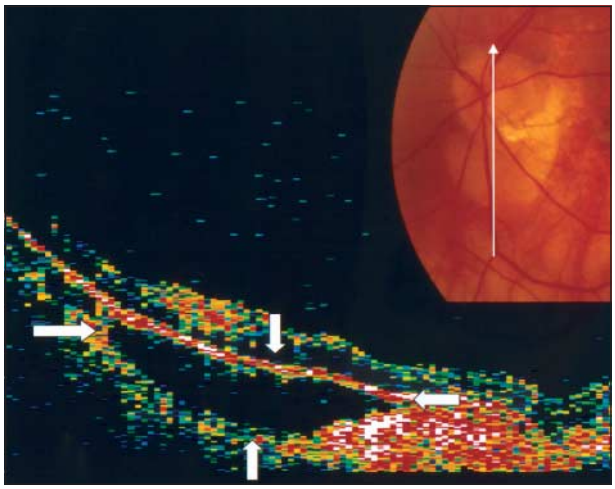

Figure 5. Case 4. Color photograph of the left eye of a 76-year-old man with pathologic myopia. There is a small, tilted disc surrounded by a myopic conus and a yellow-orange, well-demarcated elevation inferior to the disc consistent with a peripapillary detachment in pathologic myopia. An optical coherence tomographic image confirms the presence of a localized detachment that appears to involve the retina and retinal pigment epithelium (thick white arrows).

Case 6

A 41-year-old man with pathologic myopia was first examined in 1988. His visual acuity was 20/25 OU. A PDPM was evident in both eyes but it was more pronounced in the left. Optical coherence tomographic examination of the left eye re-

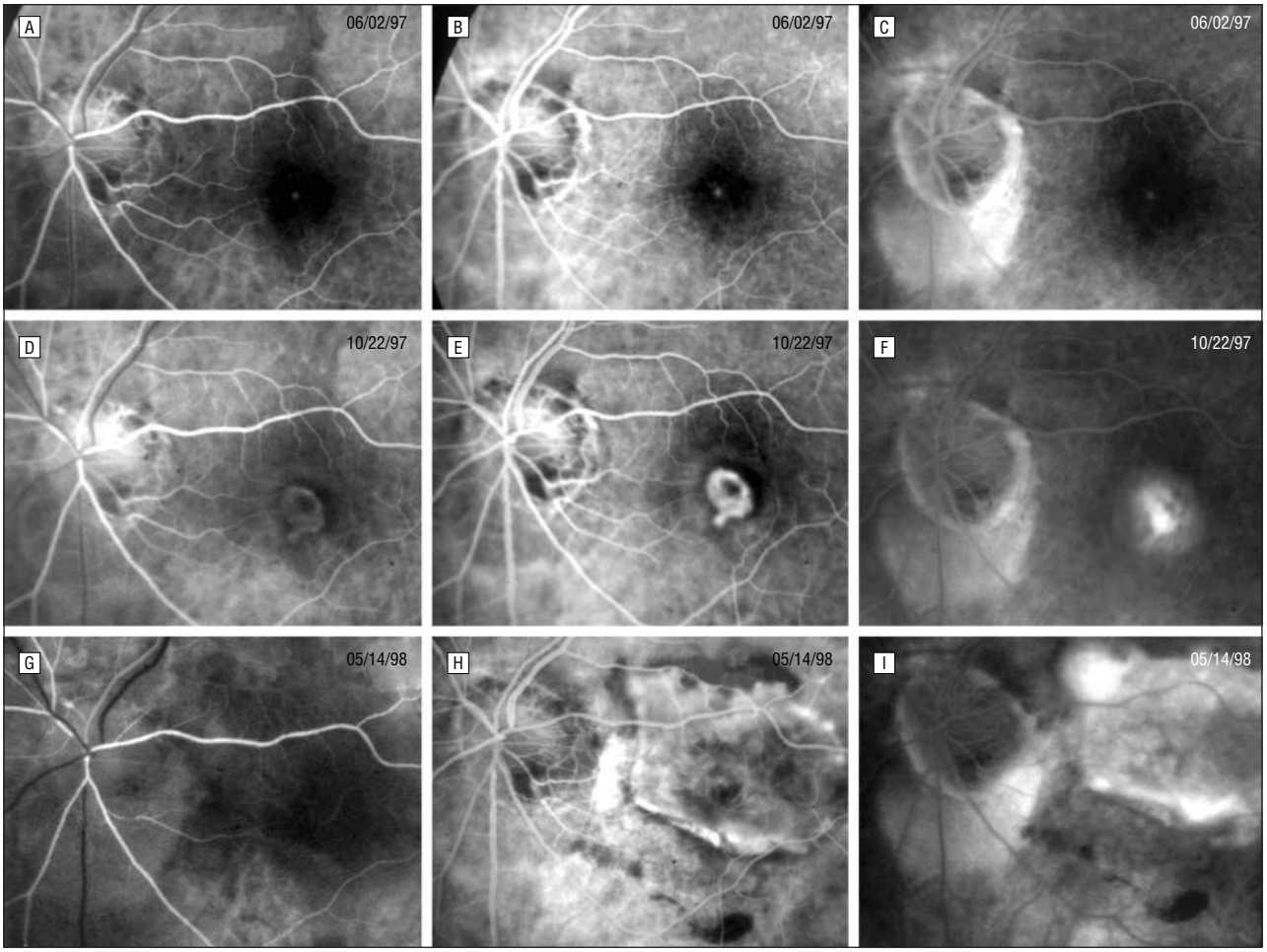

Figure 6. Case 4. Fluorescein angiogram of the left eye at initial examination (A, B, and C), 4 months later (D, E, and F), and after 1 year (G, H, and I). Early-phase fluorescein angiography (FA) study (A, D, and G) reveals relative hypofluorescence of the peripapillary detachment in pathologic myopia (PDPM). Late-phase FA study (C, F, and I) shows staining of the PDPM. There was no change in the angiographic appearance of the PDPM during this period. However, the subfoveal choroidal neovascularization progressed to the formation of a disciform scar.

vealed the presence of a localized elevation, which appeared to involve the RPE (**Figure 9**). After 10 years, there were no changes in the PDPM in either eye.

Case 7

A 43-year-old man was referred for laser photocoagulation of extrafoveal CNV in the left eye. The mean spherical equivalent correction was –13.00 D OD, and –16.00 D OS. His best-corrected visual acuity was 20/40 OD and 20/80 OS. In the right eye, there was a small tilted disc, a myopic conus, and a yellow-orange PDPM lesion inferior to the optic disc. In the left eye, there was a large area of peripapillary chorioretinal atrophy and extrafoveal CNV that was photocoagulated. A yellow-orange PDPM was present at the inferior edge of the myopic conus. During the 15 years the patient was followed up, we observed extension of the peripapillary atrophy into the PDPM in both eyes (**Figure 10**).

RESULTS

Fifteen patients (7 men and 8 women) with PDPM were identified during a 17-year period (**Table**). Five patients had bilateral involvement, for a total of 20 eyes. The mean age was 58 years (range, 40-76 years). Twelve patients were white, 2 were Asian, and 1 was Hispanic. The mean follow-up was 6 years (range, 1-15 years). The mean spherical equivalent correction was –11.00 D (range, –6.00 D to –16.00 D). The mean visual acuity at initial examination was 20/70 and the mean visual acuity at the last follow-up examination was 20/100. The median visual acuity at initial examination was 20/25. Fourteen of 20 eyes had best-corrected visual acuity of 20/40 or better at the first visit. Only 1 of these 14 eyes experienced a loss of visual acuity (from 20/30 to 20/400) secondary to the new occurrence of a choroidal neovascularization in the macula. The presence of a PDPM did not seem to affect the visual acuity. Vision loss was caused by myopic macular degeneration in 6 of 20 eyes. All patients had associated fundus changes characteristic of severe myopia. A tilted disc was present in 15 of 20 eyes, all eyes had a myopic conus, and a posterior staphyloma was present in 18 of 20 eyes. A-scan axial length measurements were obtained for 19 of 20 eyes with PDPM; the mean axial length was 27.4 mm (range, 25.3-28.9 mm).

Biomicroscopic examination of the posterior pole demonstrated the presence of a localized, well-circumscribed,

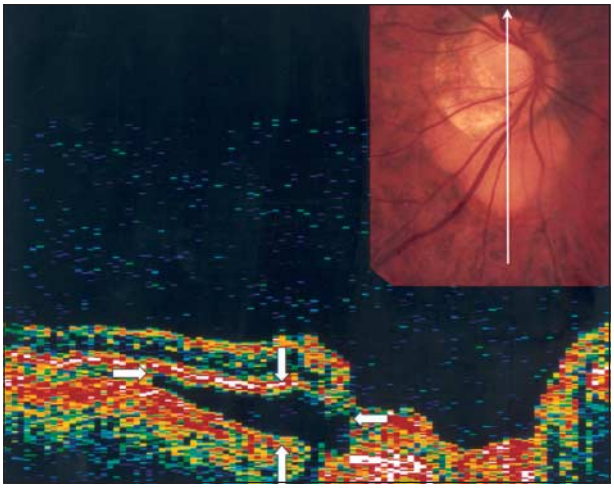

Figure 7. Case 5. Clinical photograph and optical coherence tomographic (OCT) image of the right eye of a 62-year-old man with pathologic myopia. There is a small, tilted disc, a temporal conus, and an inferior shallow yellow-orange elevation consistent with a peripapillary detachment in pathologic myopia. The OCT image demonstrates the presence of a well-circumscribed area of elevation that appears to involve both the neurosensory retina and retinal pigment epithelium (thick white arrows).

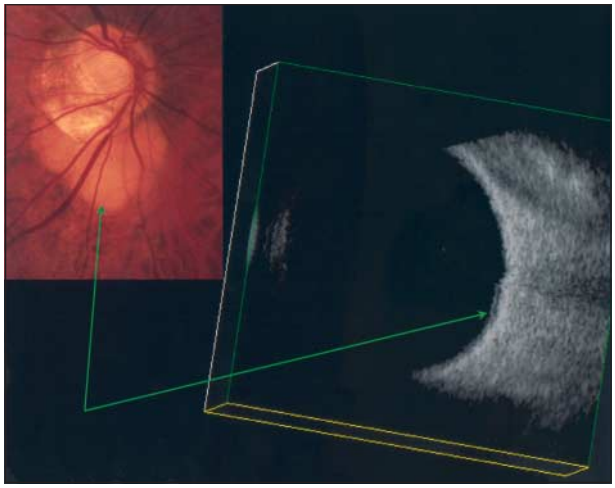

Figure 8. Case 5. A 3-dimensional B-scan ultrasound examination reveals the presence of a localized detachment corresponding to the peripapillary detachment in pathologic myopia in the same patient (arrows).

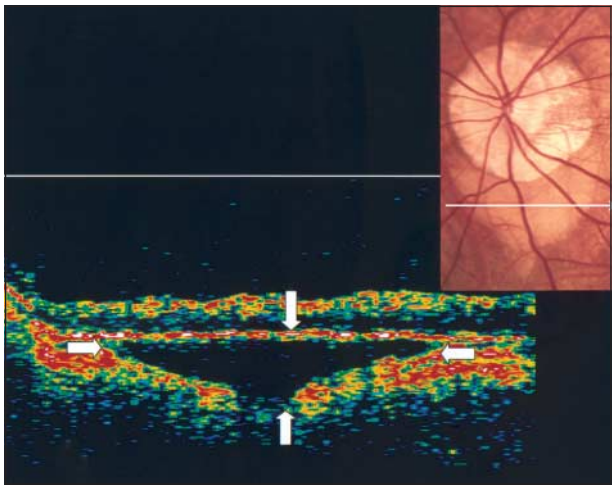

Figure 9. Case 6. Clinical photograph and optical coherence tomographic (OCT) image of the left eye. The OCT through the peripapillary detachment in pathologic myopia reveals a localized pigment epithelium detachment apparently involving the retinal pigment epithelium (thick white arrows).

yellow-orange, 1– to 2–disc diameter elevation inferior to the optic disc myopic conus. The yellow-orange lesion appeared to originate at the inferior edge of the myopic conus. It was either semicircular or triangular, with the apex pointing inferiorly. By stereoscopic examination, the lesion appeared to be a shallow detachment that was difficult to localize to either the sub-RPE or subretinal space. No additional exudative changes, such as hemorrhage or lipid exudation, were seen in association with the PDPM in any patient.

In each patient, the fluorescein angiogram showed early hypofluorescence and late staining in the area of the PDPM. There was neither early hyperfluorescence typical of serous pigment epithelial detachments (PEDs) nor leakage suggestive of active choroidal neovascularization.

In each patient, the OCT scan across the PDPM showed what appeared to be a localized detachment adjacent to the optic nerve. In all patients, the fluid appeared to be beneath both the neurosensory retina and the RPE. In 2 patients, OCT revealed an apparent discontinuity or cleft in the retinal layers at the point of transition from the PDPM to the myopic conus.

No peripapillary visual field defects were noted in 4 patients who had formal visual field testing (Humphrey 30-2). Four patients also had microperimetric examination of the area of PDPM with the scanning laser ophthalmoscope, and 3 patients had similar results on microperimetric examination with the use of the laser-aiming beam. In all patients, there were no visual field defects, either relative or absolute, detected in the area of PDPM.

During the follow-up period, no changes were noted in the size, shape, and height of the elevation, with the exception of 1 patient who had bilateral involvement. In this patient, during a 15-year follow-up, a progressive involution of the PDPM was noted in both eyes. The elevation became smaller in both eyes as peripapillary atrophy progressed into the lesion.

COMMENT

We described a series of patients with pathologic myopia and a newly recognized fundus lesion, PDPM, which does not appear similar to any other known anomaly of the myopic eye. In general, the pathogenesis of the degenerative changes at the posterior pole of the myopic eye is not well understood, although the lesions are thought to be the result of excessive axial elongation.[7] The progressive distention of the posterior pole may stretch the ocular coats, as evidenced by the straightening of the temporal retinal vessels, the appearance of a tractional myopic conus, and thinning of retina and choroid. Such signs are particularly evident in the presence of a posterior staphyloma, which represents an area of steep elongation of the globe.[8]

Histopathologic studies demonstrate the presence of characteristic changes in pathologic myopia. The sclera is thinned, particularly in the area of the posterior staphyloma, with thinning of the collagen bundles, reduced di-

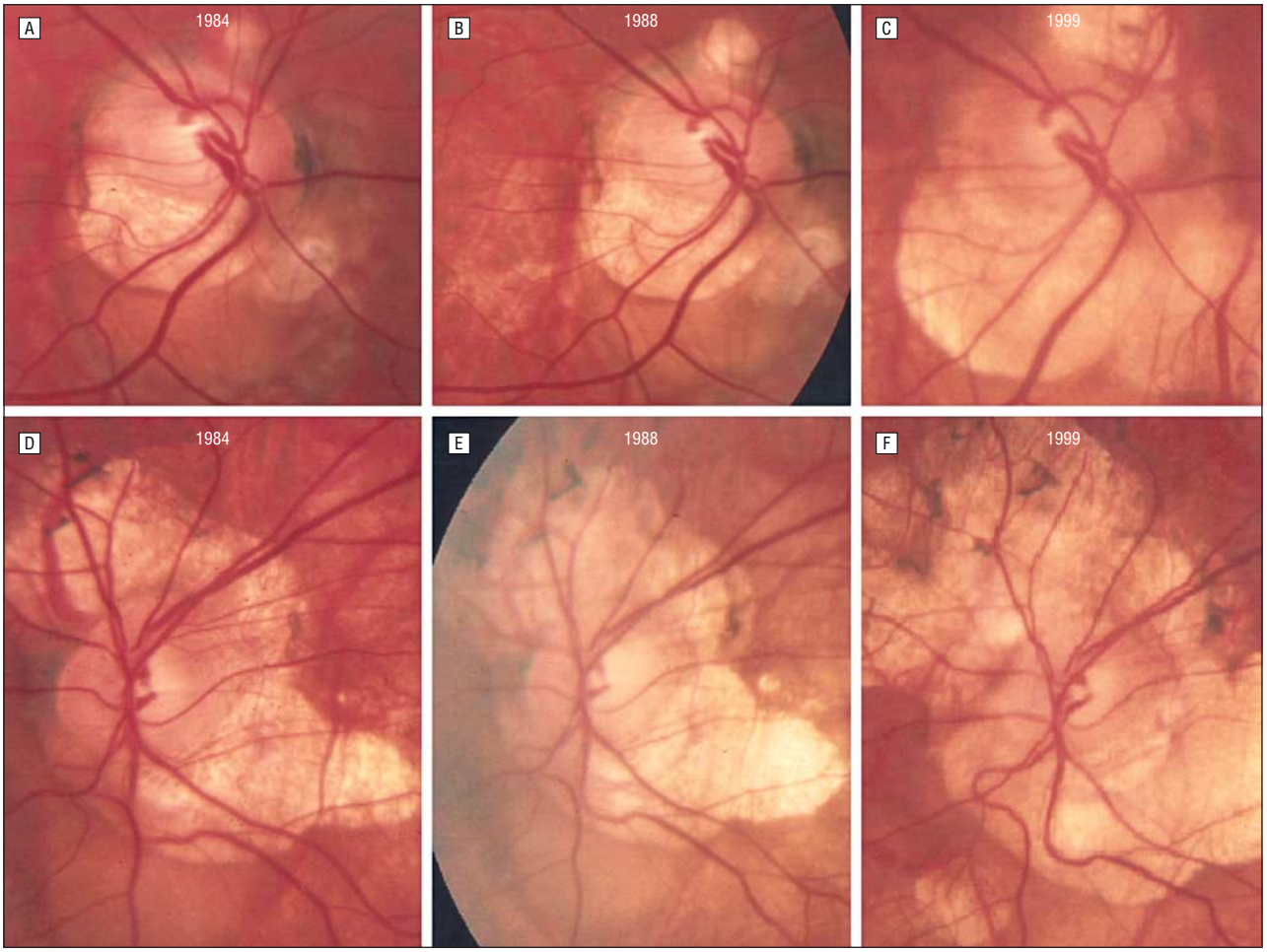

Figure 10. Case 7. Sequential clinical photographs of right (top) and left (bottom) eyes of a 43-year-old man during 15 years of follow-up. Peripapillary atrophy progressed into the peripapillary detachment in pathologic myopia lesions of both eyes, causing partial involution of the lesions.

ameter of collagen fibrils, loss of striations, and diminished scleral lamellae.[9] The choroid is thin, with a lack of vessels in some areas and a loss of choroidal melanocytes.[10] The RPE cells appear flatter and larger then usual, before their degeneration.[11] The Bruch membrane undergoes a variety of changes, including thinning, splitting and rupturing.[12]

Typical changes of the myopic disc include tilting with the temporal side flattened and the nasal side elevated with the edge raised. In addition, a concentric area of depigmentation, known as the myopic conus or temporal crescent, often surrounds the temporal side. The conus surrounds the entire disc in 10% of cases. In some cases, it extends into the macular region.[2] Finally, the presence of a posterior staphyloma is typical of degenerative myopia.[9] The posterior staphyloma is a localized ectasia of sclera, choroid, and RPE.

To our knowledge, this is the first report of what appears to be a distinct entity seen in patients who all had pathologic myopic fundus features. The PDPM lesions all shared characteristic morphologic traits. In each case, there was a localized area of elevation inferior to the optic disc and adjacent to the myopic conus. All of the lesions were yellow-orange and well circumscribed, with sharp margins. When viewed stereoscopically, the lesion appeared to be a localized detachment. However, it was impossible to distinguish between an elevation at the level of the subneurosensory or sub-RPE space. Since the lesions were semiopaque, obscuring underlying choroidal markings, we felt that the lesions were unlikely to represent a localized retinoschisis. In all cases, the OTC results seemed to localize the detachment to the sub-RPE space.

The cause of the formation of PDPM in myopic eyes is unknown. Even the name is merely descriptive. In the absence of clinicopathologic correlation, we can only speculate on the pathogenesis and nature of PDPM. While the results of OCT of the lesions appeared consistent with an RPE detachment, the fluorescein angiograms did not reveal early hypofluorescence typical of serous PEDs. Similarly, serous PEDs are almost never seen in the pathologically myopic fundus and rarely remain stable in size, shape, and height as noted in this group of patients followed up for a mean of 6 years. The fluorescein angiograms in these patients revealed more of a shallow neurosensory detachment with late staining of turbid subretinal fluid.

The consistent location of the detachment at the inferior border of the disc may be related to the early development of the eye and could suggest that the lesions are an incomplete form of choroidal coloboma. Coloboma formation results from incomplete closure of the

Demographic Data of Patients With Peripapillary Detachment in Pathologic Myopia (PDPM)*

Patient/ Sex/Age, y	Race	Follow-up, mo	SE		Initial VA		Final VA		PDPM		TD		MC		Staphyloma		AL	
			OD	OS	OD	OS	OD	OS	OD	OS	OD	OS	OD	OS	OD	OS	OD	OS
1/F/73	White	12	SE –16.00	SE –15.00	20/25	20/20	20/25	20/20	No	Yes	Yes	Yes	Yes	Yes	Yes	Yes	28.5	28.4
2/M/71	White	12	SE –6.00	SE –2.00	20/50	20/80	20/40	20/40	Yes	No	Yes	No	Yes	No	Yes	No	26.5	24.5
3/M/60	White	60	SE –16.00	SE –9.25	FC	20/20	FC	20/20	Yes	Yes	No	No	Yes	Yes	Yes	Yes	28	26.5
4/M/76	White	45	SE –10.00	SE –10.00	20/400	20/30	20/400	20/400	No	Yes	Yes	Yes	Yes	Yes	Yes	Yes	26	26.1
5/M/62	White	31	SE –8.00	SE –8.00	20/20	20/60	20/20	20/60	Yes	No	Yes	No	Yes	Yes	No	No	25.3	25.4
6/M/41	White	121	SE –8.50	SE –9.75	20/25	20/25	20/25	20/25	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	27.5	27.4
7/M/43	White	180	SE –13.00	SE –16.00	20/40	20/80	20/20	20/100	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	28	28
8/F/39	Asian	50	SE –15.00	SE –11.50	20/80	20/25	20/50	20/25	No	Yes	Yes	Yes	Yes	Yes	Yes	Yes	27.3	27.1
9/M/41	Asian	136	SE –9.00	SE –10.00	20/20	20/50	20/30	20/25	Yes	Yes	No	No	Yes	Yes	Yes	Yes	28.7	28.9
10/F/58	White	15	SE –6.00	SE –13.00	20/30	20/400	20/30	20/400	No	Yes	No	No	Yes	Yes	Yes	Yes	26	27.8
11/F/72	White	31	SE –10.00	SE –10.00	20/40	20/40	20/30	20/30	Yes	No	Yes	No	Yes	No	Yes	No	27	24
12/F/65	Latino	36	SE –8.00	SE –6.00	20/30	20/30	20/25	20/25	Yes	No	Yes	Yes	Yes	No	Yes	No	27	26
13/F/40	White	89	SE –11.00	SE –15.00	20/30	20/30	20/30	20/30	No	Yes	Yes	Yes	Yes	Yes	No	Yes	26.8	27.2
14/F/55	White	141	SE –14.00	SE –12.00	20/30	20/30	20/25	20/25	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	28	27.5
15/F/73	White	36	SE –11.00	SE –11.00	20/25	20/20	20/25	20/20	No	Yes	Yes	Yes	Yes	Yes	No	No	N/A	N/A

Abbreviations: AL, axial length; MC, myopic crescent; SE, spherical equivalent; TD, tilted disc; VA, visual acuity.

fetal fissure. The fetal fissure closes first in the region of the equator, with progressive closure anteriorly and posteriorly from that point. Failure of the fissure to close posteriorly results in the absence of choroid, pigment epithelium, and retina in that area.[13] Often, there is thinning and ectasia of the sclera. However, this did not appear to be the case in our patients. Although the lesions were located inferiorly, there was no evidence in any case of a uveal coloboma.

Another possible explanation for the consistent inferior location of the PDPM in relationship to the optic nerve is that the PDPM may simply represent gravitation of subretinal fluid originating from the area of the optic disc. The fluid could originate from the optic canal and/or the vitreous cavity. In fact, 2 patients in this series had thinning or absence of all the retinal layers at the margin of the PDPM, which were detected by scanning the edge of the lesion at an oblique angle during OCT. It is possible that other patients had similar full-thickness retinal defects that were not detected. When examined clinically, several patients appeared to have a discontinuity of the inner retinal layers at the edge of the PDPM. However, this was a subjective observation that was impossible to capture in clinical photographs. Possibly, vitreous fluid could penetrate into the subretinal space through a defect in the retina layers at the inferior margin of the myopic conus or in the area within the very atrophic conus. If this hypothesis is true, it is more probable that the PDPM represents a neurosensory detachment than a serous PED.

A similar pathogenesis has been suggested in cases of optic disc pits. Optic disc pits are dark gray excavations of the optic nerve head that may be associated with a neurosensory macular detachment and retinoschisis. Although the exact mechanism that causes the neurosensory detachment in the presence of optic disc pits is unknown, it has been postulated that vitreous fluid collects under the inner retina at the temporal edge of the pit, lifting it and the nerve fiber layer away from the outer retina and causing a macular retinoschisis. A neurosensory detachment may form subsequently, secondary to a hole in the outer retinal layers.[14]

There are similarities between optic pits and PDPM. In both cases, there is peripapillary chorioretinal atrophy, RPE changes, and localized retinal elevation in proximity to the optic disc. However, the detachments related to optic pits are usually larger, vary in size and shape, and rarely stain on fluorescein angiographic examination. Patients with optic pit–related macular retinoschisis and/or detachment usually have reduced visual acuity and may develop an outer-layer macular hole. None of the patients in our series had optic nerve findings suggestive of an optic pit. In all of our patients, the PDPM spared the fovea, remained stable in size, and did not appear to affect visual acuity.

Other optic disc anomalies are easily distinguishable from PDPM. Optic disc drusen, morning glory syndrome, optic disc hypoplasia, and dysplastic discs may show some vague resemblance to PDPM, but none of these conditions have well-demarcated areas of retina/RPE elevation like those observed in the patients in our series.[15,16]

Malignancies could also be potentially confused with PDPM. Metastatic choroidal tumors often initially appear as yellow-white elevations, deep to the retina. However, metastatic choroidal tumors are usually larger than PDPM, are often multifocal, and do not have a predilection for the peripapillary area or the myopic eye. When present, the exudative detachment associated with choroidal metastasis tends to be larger than PDPM and tends to fluctuate in size.[17] None of our patients had a positive history for systemic malignancies. Furthermore, the stability of the PDPM in our patients argues strongly against a metastatic lesion.

Finally, PDPM needs to be differentiated from other causes of serous PED and/or neurosensory retina, such as myopic and age-related macular degeneration, polypoidal choroidal vasculopathy (polypoidal choroidal neovascularization), peripapillary choroidal neovascularization, and central serous chorioretinopathy.[18]

In our series, 9 of 15 patients were older than 50 years, and 7 of 15 patients had some evidence of choroidal neovascularization secondary to pathologic myopia in the central macula. None of the patients had drusen

typical of age-related macular degeneration. Furthermore, PEDs are almost never seen in the myopic fundus in association with age-related/myopic macular degeneration. Hemorrhage, a common feature of neovascularized maculopathies, was not seen in association with any of the PDPM lesions. The presence of early hypofluorescence seen on fluorescein angiographic examination of the PDPM lesions also argued against CNV. Last, the stability of PDPM over time also suggested that these lesions did not represent CNV.

Polypoidal choroidal vasculopathy may be a cause of serous/hemorrhagic juxtapapillary PEDs and neurosensory detachment. Polypoidal choroidal vasculopathy tends to occur more commonly in highly pigmented fundi with no other abnormalities.[19,20] However, dilated choroidal vascular channels and subretinal/sub-RPE hemorrhages are a frequent early manifestation of polypoidal choroidal vasculopathy. None of our patients had any evidence of choroidal vascular abnormalities and of subretinal hemorrhages associated with PDPM.

In central serous chorioretinopathy, there is formation of a serous PED and of an overlying larger neurosensory detachment. In some severe cases of central serous chorioretinopathy, there is also deposition of a whitish material consistent with fibrin under the PED.[21] However, the fluorescein angiographic examination in central serous chorioretinopathy demonstrated leakage of dye through a defect in the RPE into the subretinal space. This is different from what is seen in PDPM where there is early blockage of fluorescence and late staining of the lesion. In age-related macular degeneration and central serous chorioretinopathy, the PEDs tend to evolve over time either toward progression and consolidation into a disciform scar (age-related macular degeneration) or toward resolution (central serous chorioretinopathy). In 13 of 14 patients in this series, there were no changes in the appearance of the PDPM during the extended follow-up period. In 1 patient, there was partial resolution of the PDPM during a 15-year follow-up.

In conclusion, we report for the first time, to our knowledge, on an asymptomatic, yellow-orange, peripapillary lesion that clinically appeared to be a localized detachment of the neurosensory retina or RPE. All patients had fundus changes consistent with pathologic myopia. The results of OCT evaluations of these eyes confirmed the presence of a localized detachment that appeared to be at the level of the RPE. These lesions were nonprogressive and not associated with visual changes during the follow-up period. Recognition of this lesion as a separate entity associated with pathologic myopia is important so that it is not confused with other fundus abnormalities, such as tumors or CNV, which necessitate further investigation and treatment.

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