Peripapillary Staphyloma

Clinical Features and Visual Outcome in 19 Cases

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Objective: To investigate the clinical features and visual outcome of patients with peripapillary staphyloma.

Methods: The medical records of patients diagnosed as having peripapillary staphyloma at Seoul National University Children's Hospital, Seoul, Korea, between January 1, 1991, and December 31, 2003, were reviewed. Visual acuity and associated ocular and systemic abnormalities were recorded.

Results: Nineteen patients (21 eyes) with peripapillary staphyloma were included in the study. The mean age at initial examination was 21 months, and 42% (8/19) of the patients were girls. Two patients (11%) had bilateral peripapillary staphylomas. Only 2 eyes had visual acuity better than 20/200 at the last examination, and 7 eyes had severe myopia of more than −6 diopters. After occlusion therapy, 1 patient achieved a visual improvement from light perception to 20/30. Congenital cataract and persistent pupillary membrane were present in 2 eyes each at initial examination. Complications observed during the follow-up period included acquired total cataract in 2 eyes and retinal detachment in 3 eyes. Scleral encircling buckling was performed in 1 eye with retinal detachment, and subsequently the retina was reattached. Only 2 patients had accompanying systemic abnormalities.

Conclusions: Rarely, patients with peripapillary staphyloma can achieve significant visual improvement by occlusion therapy, although visual outcome is generally poor. Because associated ocular disease and refractive errors are not infrequent, complete ophthalmic examinations and regular follow-up are necessary.

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PERIPAPILLARY STAPHYLOMA IS A rare nonhereditary congenital optic disc anomaly, in which a deep fundus excavation surrounds a relatively normal-appearing optic nerve head.1,2 It is generally unilateral and is accompanied by poor vision, although a bilateral case with normal visual acuity has been reported.3 Unlike other excavated optic disc anomalies, peripapillary staphyloma is known to be rarely associated with other congenital defects or systemic diseases.2

Its exact pathophysiology is unknown. However, the clinical features of peripapillary staphyloma are thought to be most consistent with diminished peripapillary structural support, resulting from incomplete differentiation of the posterior sclera from neural crest cells in the fifth month of gestation.4

A few case reports of peripapillary staphyloma have been published. However, as far as we know, no report has been published on a large series. Moreover, it is not known whether occlusion therapy is helpful in patients with peripapillary staphyloma. In this study, we investigated the clinical features, effectiveness of occlusion therapy, and visual outcome of 19 patients with peripapillary staphyloma.

METHODS

The medical records of 19 patients (21 eyes) with peripapillary staphyloma who were diagnosed at Seoul National University Children's Hospital, Seoul, Korea, between January 1, 1991, and December 31, 2003, were reviewed. The diagnosis of peripapillary staphyloma was based on (1) deep fundus excavation surrounding a relatively normal-appearing disc, (2) a normal retinal vasculature pattern, (3) no central glial tuft on the disc, and (4) no iris or retinochoroidal coloboma.

The chief complaint at initial examination, family history, and details of systemic reviews were investigated. Measurement of uncorrected visual acuity and best-corrected visual acuity (BCVA), fixation testing in case of preverbal age (in patients older than 3 months),
The Krimsky test, alternative prism cover and uncover test if possible, examination of ductions and versions, cycloplegic refraction, slitlamp examination, and fundus examination were all undertaken. Ultrasonography was performed to evaluate the depth and the width of excavation.

Patch occlusion therapy with full refractive correction was routinely advocated to treat possible accompanying functional amblyopia except in cases of no light perception on reliable examination, bilateral involvement, the coexistence of another severe ophthalmic abnormality, and better visual function than in the contralateral eye. Part-time occlusion (2 to 6 h/d) was performed to avoid reverse amblyopia.

RESULTS

Clinical features are described in the Table. The mean age at initial examination was 21 months (range, 1-78 months), and the mean follow-up period was 58 months (range, 6-152 months). Eight (42%) of the patients were girls. Two patients (11%) had bilateral peripapillary staphylomas. All patients were Asians. No patient had a family history of optic disc anomaly. Of the chief complaints at initial examination, poor vision or poor fixation (8 patients) was most frequent, followed by strabismus (4 patients), nystagmus (3 patients), small eyeball (1 patient), iris abnormality (1 patient), head turn (1 patient), and an incidental finding of a contralateral small eyeball (1 patient).

Visual acuities at first and last examinations are shown in the Table. Only 2 eyes had visual acuity better than 20/200 at the last examination. Seven of the 21 eyes had a severe myopia of more than −6 diopters (D), although none had an excavation involving the macula. Occlusion therapy (2 to 6 h/d) was performed in 13 eyes. Of 6 patients in whom occlusion therapy was not performed, 2 had bilateral peripapillary staphylomas, 1 had associated severe microphthalmia, 1 had no light perception at initial examination, 1 had contralateral microphthal-
Occlusion therapy was initiated at a mean age of 19 months (range, 5-78 months), and the mean duration of therapy was 30 months (range, 12-78 months). After occlusion therapy, no one achieved the same visual acuity in the affected eye as in the sound eye. However, 1 patient (case 17, Figure 1) achieved significant visual improvement. He was able to fix and follow light at 4 months of age. After starting occlusion therapy for 2 hours per day at 6 months of age, he showed good compliance. After 32 months of occlusion, at which time the Snellen chart can be used to examine visual acuity reliably, the BCVA of the affected eye was 20/100 and that of the sound eye was 20/25. Since then, his BCVA gradually improved with occlusion therapy for 4 hours per day. At the patient’s last examination, at 7 years of age, the BCVA in his affected eye was 20/30 and occlusion therapy was maintained for 6 h/d.

Contraction of peripapillary staphyloma was not found in any patient despite prolonged examination and light provocation of the contralateral eye. The depth and the width of the peripapillary staphyloma were highly variable. The mean depth was 4.61 mm (range, 1.07-9.92), and mean width, 5.34 mm (range, 2.40-10.52) (Figure 2).

Microphthalmia was detected in 1 patient and congenital cataract was present in 2 patients at initial examination. A persistent pupillary membrane was found in 2 patients, persistent hyperplastic primary vitreous in 1, contralateral severe microphthalmia in 1, and contralateral morning glory disc anomaly in 1. During the follow-up period, acquired total cataract developed in 2 patients. Of the 2, 1 eye (the left eye of patient 2) had had no cataract at initial portable slitlamp examination, and total cataract was detected at the age of 31 months. The other eye (in patient 7) had anterior polar cataract at ini-
tial examination that progressed to total cataract by the age of 67 months. These were treated by lensectomy. However, visual acuity was not improved after surgery. Five patients developed significant misalignment of the eyeballs, and 3 of these underwent strabismus surgery.

Three of 21 eyes developed total retinal detachment without a retinal break during the follow-up period. Retinal detachment was detected by routine examination, and the onset could not be identified. Surgery was advised in all patients. One patient (case 3), whose BCVA had once been 12/200 OD, had no light perception in the right eye at the age of 11 years, and the parents refused surgery. The second (case 2, Figure 3), in whom the retinal detachment was detected at age 21 months, underwent scleral encircling buckling with retinal reattachment afterward. The third (case 6), whose BCVA had once been 20/200, underwent a vitrectomy with intravitreal gas (sulfur hexafluoride, 20%) tamponade immediately after examination at the age of 72 months, and the retina was flattened. However, after a few weeks, the retina was re-detached. The parents refused reoperation.

Gestational age and body weight at birth were within the normal range in all patients. Only 2 patients had a systemic abnormality. Of these, 1 had a developmental delay and the other had a median cleft, which was treated by cheiloplasty.

**COMMENT**

This series represents, to our knowledge, the largest reported study of peripapillary staphyloma. Moreover, we report what we believe to be the first case of peripapillary staphyloma, in which visual acuity was improved after conventional occlusion therapy, and the first case where total retinal detachment was a complication of peripapillary staphyloma and was successfully treated with a scleral encircling buckling procedure.

Peripapillary staphyloma is often confused with other excavated congenital optic disc anomalies. In optic disc coloboma, the optic disc contains the excavation. Iris or retina coloboma often accompanies it, and systemic dis-
case is also frequently present. In morning glory disc anomaly, excavation is usually shallower than in peripapillary staphyloma, and there is a centrally located peripapillary glial tuft. In addition, an anomalously enlarged optic disc and increased, straightened vessels radiating outward are found. In this study, we adopted strict diagnostic criteria to exclude the morphologically similar excavated optic disc anomaly.

In the present study, visual outcome was generally poor and mainly ranged from 20/200 to counting fingers, which is compatible with the findings of a previous report, but we also documented cases with BCVA of 20/30 and of no light perception. It is known that patients with peripapillary staphyloma are usually emmetropic or slightly myopic, and the findings of the present study concur. However, in contrast to previous reports, 7 (33%) of 21 eyes had severe myopia of more than −6 D, and 5 of these had severe myopia of more than −10 D. In addition, 2 of those with myopia of more than −6 D had BCVA of 20/100 and 20/30, respectively, at the final examination. Therefore, early refraction is necessary to correct refractive errors.

Kushner reported that functional amblyopia could co-exist with structural optic disc abnormalities and that in such cases visual acuity could be improved by conventional occlusion therapy. Moreover, 1 case report described peripapillary staphyloma with normal vision. However, no report is available on visual outcome after occlusion therapy in patients with peripapillary staphyloma. We used occlusion therapy in 13 eyes and 1 showed possible visual improvement; this represents the first case, to our knowledge, with peripapillary staphyloma in which visual acuity improved after occlusion therapy. Some of the visual improvement might have resulted from more accurate results of vision testing being obtained at an older age, although we attribute most of the visual improvement to occlusion therapy. In most cases, poor compliance and no visual improvement have caused us to discontinue occlusion therapy prematurely. Indeed, Yang and Lambert reported that prolonged occlusion therapy might lead to significant psychosocial harm and developmental delay. Thus, although it is advisable to attempt occlusion therapy in all cases of peripapillary staphyloma, a regular vision check is mandatory and reckless, prolonged occlusion must be avoided.

Several reports have been published on contractile peripapillary staphyloma. Light stimulus to the contralateral eye might provoke contraction of the peripapillary staphyloma. In this study, we carefully examined the disc for a long time with a provoking light stimulus to the contralateral eye, but no contraction was observed in any case. In previous reports of contractile peripapillary staphyloma, the patients were generally older (age range, 15–42 years) than in our series. Therefore, it may be that our patients were too young and the neuromuscular contraction mechanism had not matured. Another possible cause is poor cooperation with respect to long-term fundus examination. In cases of poor cooperation, we sedated the patient and then performed the examination so as not to miss the peripapillary staphyloma contraction.

In previous reports, the depth of peripapillary staphylomas was highly variable, ranging up to 9.77 mm. In the present study, depths and widths were also variable, especially between the eyes of a single patient. Because of the statistically small number of cases, we cannot confirm that the depth and width of peripapillary staphyloma correlate with visual outcome.

In our study, an associated ocular abnormality was not rare. These included congenital cataract and developmental anomalies such as microphthalmia, persistent hyperplastic primary vitreous, and a persistent pupillary membrane. Moreover, acquired abnormalities such as retinal detachment or total cataract were observed as complications during the follow-up period. Therefore, not only is a complete examination of both eyes mandatory at the first visit, but regular follow-up is essential as well.

Retinal detachment can sometimes be associated with congenital optic disc anomalies, such as morning glory disc anomaly, optic disc coloboma, or congenital pit of the optic disc. A few reports on peripapillary staphyloma associated with limited retinal detachment have been published. However, total retinal detachment has not previously been reported in patients with peripapillary staphyloma, to our knowledge. In the 3 patients with retinal detachment during the follow-up period, no retinal tear had been found preoperatively or intraoperatively. The exact cause of retinal detachment associated with peripapillary staphyloma is not known. However, the pathophysiology might be similar to that of retinal detachment in combination with another excavated optic disc anomaly. Several theories have been proposed, including abnormal communication between the subarachnoid space of the optic nerve and the subretinal space, communication between the vitreous cavity and the subretinal space, and communication with the orbit through a defect in the optic nerve sheath via disruption of the intermediary tissue of Kuhnt. Further study must be undertaken to prove that this hypothesis can be applied to peripapillary staphyloma. We cannot explain the pathophysiologic mechanism that caused the retinal flattening after scleral encircling buckling. However, there is a possibility that a hidden break was located in the periphery of the retina.

This study confirms that peripapillary staphyloma is rarely associated with systemic abnormalities, although rare associations have been reported, including atypical hemimegalencephaly with hypoplasia of an optic radiation, frontonasal dysplasia with basal encephalocele, and orofacial capillary hemangioma. In the present study, 1 patient had a median cleft, but encephalocele was not found by imaging study.

In conclusion, eyes with peripapillary staphyloma may rarely achieve visual improvement by occlusion therapy, but visual outcome is generally poor. A complete ophthalmic examination and regular follow-up are necessary because associated ocular disease and refractive errors were not found to be rare.

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Financial Disclosure: None.

Additional Information: Dr Kim had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

REFERENCES


ARCHIVES Web Quiz Winner

June 2005 Web Quiz Winner

C ongratulations to the winner of our June quiz, Thabit Ali Mustafa Odat, FRCSGlasg, King Hussein Medical Center, Royal Medical Services of Jordan, Amman, Jordan. The correct answer to our June challenge was uveoparotid fever. For a complete discussion of this case, see the Photo Essay section in the July ARCHIVES (Blair MP, Rizen M. Heerfordt syndrome with internal ophthalmoplegia. Arch Ophthalmol. 2005;123:1017).

Be sure to visit the Archives of Ophthalmology Web site (http://www.archophthalmol.com) and try your hand at our Clinical Challenge Interactive Quiz. We invite visitors to make a diagnosis based on selected information from a case report or other feature scheduled to be published in the following month’s print edition of the Archives. The first visitor to e-mail our Web editors with the correct answer will be recognized in the print journal and on our Web site and will also be able to choose one of the following books published by AMA Press: Clinical Eye Atlas, Clinical Retina, or Users’ Guides to the Medical Literature.
Correspondence: Dr Goldberg, Jules Stein Eye Institute, University of California, Los Angeles, School of Medicine, 100 Stein Plaza, Los Angeles, CA 90095-7006 (goldberg@jsei.ucla.edu).

REFERENCE


Correction

Errors in Table. In the Clinical Sciences article by Kim et al titled “Peripapillary Staphyloma: Clinical Features and Visual Outcome in 19 Cases,” published in the October issue of the ARCHIVES (2005;123:1371-1376), 2 errors occurred in the Table on page 1372. In that table, the footnote symbol attached to the refraction value for the left eye of patient 2 should have indicated that the value was available preoperatively. In addition, the footnote symbol attached to the best-corrected visual acuity at the first examination for patient 6 should have indicated that because of preverbal age, visual acuity was assessed as LP (light perception) or NLP (no light perception) by fixation test. As a result of these errors, the footnotes in the published Table were out of order. The corrected Table is reprinted here in its entirety.

Table. Clinical Features and Visual Outcome of Patients With Peripapillary Staphyloma

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Sex</th>
<th>Age, mo*</th>
<th>Follow-up, mo</th>
<th>First Examination</th>
<th>Last Examination</th>
<th>Refraction (SE), D</th>
<th>Depth, mm</th>
<th>Width, mm</th>
<th>Result of Occlusion</th>
<th>Associated Abnormality</th>
<th>Complication</th>
<th>Operation</th>
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<tr>
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<td>46</td>
<td>39</td>
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<td>LP</td>
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<td>NA</td>
<td>ND</td>
<td>Contralateral MGD anomaly, developmental delay</td>
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<td>None</td>
</tr>
<tr>
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<td>M</td>
<td>46</td>
<td>39</td>
<td>NLP†</td>
<td>LP</td>
<td>−11.00</td>
<td>NA</td>
<td>NA</td>
<td>ND</td>
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<td>None</td>
</tr>
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<td>LP</td>
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<td>3.07</td>
<td>5.55</td>
<td>ND</td>
<td>RD</td>
<td>Encircling buckling</td>
<td>Lensectomy</td>
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<td>152</td>
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<td>LP</td>
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<td>Total cataract</td>
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<td>6/M/27/OS</td>
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<td>27</td>
<td>63</td>
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<tr>
<td>7/F/61/OS</td>
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<td>61</td>
<td>6</td>
<td>NLP</td>
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<td>Poor</td>
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<td>10.52</td>
<td>ND</td>
<td>Antrior polar cataract</td>
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<td>None</td>
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<td>94</td>
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<td>3.63</td>
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<td>3.84</td>
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<td>Exotropia§</td>
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<td>10/M/1/OD</td>
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<td>37</td>
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<td>CF</td>
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<td>57</td>
<td>NLP†</td>
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<td>Poor</td>
<td>5.62</td>
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<td>92</td>
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<td>20/100</td>
<td>−11.50</td>
<td>2.65</td>
<td>5.77</td>
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<td>101</td>
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<td>CF</td>
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<td>7.45</td>
<td>9.78</td>
<td>Failed</td>
<td>Median cleft</td>
<td>Exotropia§</td>
<td>Strabismus operation, cheioplasty</td>
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<td>82</td>
<td>LP†</td>
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<td>CF</td>
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<td>2.94</td>
<td>3.78</td>
<td>Failed</td>
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</table>

Abbreviations: BCVA, best corrected visual acuity; CF, counting fingers; D, diopters; LP, light perception; MGD, morning glory disc anomaly; NA, not available; ND, not done; NLP, no light perception; PHPV, persistent hyperplastic primary vitreous; PPM, persistent pupillary membrane; RD, retinal detachment; SE, spherical equivalent.

*Age at first examination.
†For these measurements, because of preverbal age, visual acuity was assessed as LP or NLP by fixation test.
‡Preoperatively available refractive value.
§Cosmetically significant misalignment of eyeballs.