Posterior Staphylomas in Pathologic Myopia Imaged by Widefield Optical Coherence Tomography

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PURPOSE. To examine posterior staphylomas by widefield optical coherence tomography (WF-OCT) and three-dimensional magnetic resonance imaging (3D-MRI).

METHODS. Highly myopic patients (myopic refractive error >8.0 diopters or axial length >26.5 mm) who had previously undergone orbital 3D-MRI were examined by WF-OCT.

RESULTS. The study included 100 eyes of 57 patients with a mean age of 67.9 ± 10.7 years (range, 44–85 years) and mean axial length of 30.0 ± 2.3 mm (range, 25.1–36.5 mm). All staphylomas detected on the 3D-MRI, except for two very large staphylomas, were visualized on the WF-OCT images. Morphologic hallmarks of the staphylomas were smoothly configured staphyloma border with a gradual thinning of the choroid and an inward protrusion of the sclera at the staphyloma edge. Comparing the detectability of the staphylomas on the WF-OCT images versus 3D-MRI revealed no significant difference between both techniques (P = 0.12, χ² test). Comparing the staphyloma classification between both techniques showed a good concordance with a concordance index kappa of 0.61 (95% confidence interval: 0.50–0.72). A spatial relationship between the staphylomatous areas and the macula and optic nerve head was observed by WF-OCT.

CONCLUSIONS. WF-OCT can provide tomographic images of posterior staphylomas in a resolution and size unachievable so far, and may replace 3D-MRI in assessing posterior staphylomas. Future studies using WF-OCT may explore the detailed morphologic characteristics of posterior staphylomas and give clues to the etiology of staphylomas.

Keywords: posterior staphyloma, pathologic myopia, swept-source optical coherence tomography, three-dimensional magnetic resonance imaging, widefield OCT

A posterior staphyloma is a hallmark of pathologic myopia,1–3 and has been defined by Spaide2 as an outpouching of the ocular wall with a curvature radius being smaller than the curvature radius of surrounding ocular wall. Although typical for pathologic myopia, posterior staphyloma also can occur in eyes that are not highly myopic.1,4,5 Previous studies revealed that highly myopic eyes with posterior staphylomas had a significantly worse visual and anatomic outcome than highly myopic eyes without staphylomas.6–8

Despite its importance, the description of posterior staphylomas has so far been based mostly on ophthalmoscopy. Based on their ophthalmoscopic appearance, Curtin5 differentiated posterior staphylomas into 10 different types in 1977. Later, Moriyama and colleagues9,10 applied three-dimensional magnetic resonance imaging (3D-MRI), and Ohno-Matsui and coworkers11,12 used a combination of 3D-MRI and ultra-widefield fundus imaging to classify posterior staphylomas into six different types, based on the size, shape, and location of the staphylomas.

Three-dimensional MRI has an advantage of visualizing the shape of the whole eye, including the anterior ocular segment. Three-dimensional MRI is, however, not feasible as a screening technique, and due to a relatively low spatial resolution, subtle changes of shallow staphylomas are difficult to detect. Also, 3D-MRI cannot differentiate among the retinal, choroidal, and scleral tissue. Because the 3D-MRI technique uses T2-weighted images showing intraocular fluid, 3D-MRI demonstrates the vitreo-retinal interface or the inner surface of the retina and does not show local variations in the thickness of the choroid and sclera.

Optical coherence tomography (OCT) analyzes the curvature of the sclera in eyes with pathologic myopia.7,13 Due to the limited scan length and depth of devices previously available, the OCT technology was limited in its usefulness for visualization of posterior staphylomas. As compared with spectral-domain OCT, the recently developed swept-source OCT technology improved the detectability of staphylomas; however, was markedly limited by its relatively short length of the scan line.14 Attempts to overcome this limitation included combining multiple scan lines in a process of photo montaging or by placing a ±20-diopter lens between the eye and the OCT device; one tried to overcome that limitation.15–21 In most of these studies, however, the widefield OCT images were restricted to one or a few scan lines, so that it was not possible to generate a three-dimensional image of staphylomas.
A new prototype of a widefield swept-source OCT system uses not only one but multiple scan lines and generates scan maps allowing the three-dimensional reconstruction of posterior staphylomas in a region of interest of $16 \times 14$ mm and a depth of 5 mm. Applying widefield OCT (WF-OCT), we conducted this study to visualize posterior staphylomas in highly myopic eyes in their full three-dimensional extent, to compare the detectability of staphylomas by WF-OCT and by 3D-MRI, and to describe OCT-based characteristics of staphylomas found particularly at the edge of a staphyloma.

**Figure 1.** New classification and renaming of staphyloma types according to their location and shape.

**Figure 2.** Wide macular staphyloma as visualized by WF-OCT and 3D-MRI. (A) Image of the left ocular fundus of a 79-year-old woman (axial length, 26.6 mm) showing the border of a wide staphyloma as indicated by pigmentary abnormalities (arrowheads). Arrows show the scanned lines of the WF-OCT images shown in (D) and (E). (B, C) Three-dimensional MRI viewed from the inferior (B) and from the nasal side (C), showing a wide macular staphyloma (arrowheads) with a notch at the temporal border (arrow in [B]). (D, E) Cross-sectional WF-OCT images. (D) Horizontal scan. (E) Vertical scan. An inward protrusion of the sclera and a thinning of the choroid are observed at the edge of the staphyloma in the horizontal section and in the vertical section (arrow). The staphylomatous region shows a posterior displacement of the sclera nasal to the staphyloma edge in (D) and inferior to the staphyloma edge in (E). (F–H) Three-dimensional WF-OCT images viewed from the anterior (F), the nasal (G), and from the inferior side (H), show a scleral outpouching (arrowheads) due to a wide macular staphyloma. In (F), the staphyloma border is spatially associated with the optic nerve head and the retinal vessels.
METHODS

According to the tenets of the Declaration of Helsinki, the study was approved by the Ethics Committee of Tokyo Medical and Dental University and all study participants signed an informed consent. The study included consecutive highly myopic patients who had previously undergone a 3D-MRI for the assessment of the eye shape in the primary gaze position. The patients were prospectively examined by WF-OCT between December 2015 and June 2016, using a new prototype of OCT device. Inclusion criteria for the study were high myopia, defined as a myopic refractive error (spherical equivalent) of more than \(8.0\) diopters or an axial length greater than \(26.5\) mm, and an examination by 3D-MRI for the examination of the eye shape within the past 2 years. Even eyes with a myopic refractive error less than \(8.0\) diopters were included if their axial length was greater than \(26.5\) mm. Exclusion criteria were OCT images of poor quality, mainly due to media opacities such as dense cataract, and a history of vitreoretinal surgeries or glaucoma surgeries that could have affected the shape of the globe.

All patients underwent a comprehensive ophthalmologic examination including refractometry, ocular biometry (IOL Master; Carl Zeiss Meditec, Jena, Germany), stereoscopic fundus examination in medical mydriasis, and color fundus photography (TRC-50DX, Topcon, Tokyo, Japan; or Optos 200Tx’s scanning laser ophthalmoscope; Optos PLC, Dunfermline, Scotland, UK).

The 3D-MRI examination of the globes was performed in primary gaze position as described in detail previously. The MRI scanner (Signa HDxt 1.5T, version 15; General Electric Healthcare, Waukesha, WI, USA) used a fat-suppressed T2-weighted cube as an improved sequence of a three-dimensional fast-spin-echo (256×320×256 matrix, 22-cm field of view, 1.2-mm slice thickness, repetition time 250 ms, echo time 90 ms, echo train length 90). Volume renderings of the images were generated from high-resolution three-dimensional data on a computer workstation (v. AW 4.4; GE Healthcare). The margins of the globes were detected semiautomatically by differences in signal intensity. The images of the tissues surrounding the globe were removed.

For the widefield swept-source OCT (WF-OCT), a prototype device (Canon Corp., Tokyo, Japan) was used that had an A-scan repetition rate of 100,000 Hz. The light source was a wavelength tunable laser centered at 1050 nm with a 100-nm tuning range. The scan line length was 16 mm in the horizontal direction and 14 mm in the vertical direction, and the scan depth was 5 mm. Cross scans centered on the fovea and map scans centered on the midpoint between the fovea and the optic disc were obtained. The map scan was performed with...
256 horizontal single scans in the specified area. The three-dimensional images of the posterior globe were reconstructed from the map scans by ImageJ software (https://imagej.nih.gov/ij; provided in the public domain by the National Institutes of Health, Bethesda, MD, USA). The time for image acquisition both for the cross scan and the three-dimensional scan was less than 4 seconds. As recently described by Spaide,2 we defined posterior staphylomas as an outpouching of the ocular wall with a curvature radius that was smaller than the curvature radius of the surrounding ocular wall. As described in the previous study, the posterior staphylomas were classified into six types: the wide macular type, the narrow macular type, the peripapillary type, the nasal type, the inferior type, and others (Fig. 1).11 The presence and types of staphylomas as examined by 3D-MRI and by WF-OCT were determined by two experienced examiners (KS, KO-M) independently of each other.

The statistical analysis was carried out using the program Prism 5.0c (GraphPad Software, Inc., San Diego, CA, USA). The detectability of staphylomas was compared between 3D-MRI and WF-OCT by the $\chi^2$ test. Concordance between the findings obtained on the 3D-MRI and on the WF-OCT images was estimated by calculating Cohen’s kappa coefficient ($\kappa$) with the kappa function. We considered a $\kappa$ value $\geq0.8$ as excellent concordance and a $\kappa$ value between 0.6 and 0.8 as good concordance. A $P$ value $<0.05$ was considered statistically significant.

**RESULTS**

The study included 104 eyes of 57 patients whose eye shape had previously been examined by 3D-MRI and who had additionally undergone WF-OCT. Four eyes were excluded due to poor quality of the OCT images (two eyes) or due to a history of scleral buckling surgery (two eyes). Eventually 100
eyes of 56 patients (12 men, 44 women) were enrolled with a mean age of 67.9 ± 10.7 years (range, 44–85 years), a mean refractive error (spherical equivalent) of −4.1 diopters (range, −4.5 to −21.0 diopters) and a mean axial length of 30.0 ± 2.3 mm (range, 25.1–36.5 mm).

On the WF-OCT images, all staphylomas with the exception of two large staphylomas that were detected by 3D-MRI were located in their whole length and width within the scanned region of interest (Figs. 2–5). Morphologic hallmarks of the posterior staphylomas as examined by WF-OCT were a smoothly configured border with a gradual thinning of the choroid from the periphery toward the edge of the staphyloma and a gradual rethickening of the choroid in direction toward the posterior pole. Additionally, there was a gradual thickening and inward protrusion of the sclera at the staphyloma edge. The three-dimensional WF-OCT images also visualized the spatial relationships between the staphyloma and the optic nerve head as well as retinal vessels (Figs. 2–5). Layers of vitreous were observed in some cases without large liquefaction of vitreous. In the eyes without posterior staphylomas, OCT features indicating a staphyloma edge were not detected, although the scleral curvature deviated from an ideal circular shape due to the axial elongation of the globe (Fig. 6).

Comparing the detectability of the staphyloma on the WF-OCT images with 3D-MRI revealed that the staphylomas were detected by WF-OCT in 75 of the 100 eyes and by 3D-MRI in 65 of the 100 eyes, with no statistically significant difference between both techniques in the frequency of staphyloma detection ($P = 0.12; \chi^2$ test). Among 35 eyes in which staphylomas were not detected by 3D-MRI, staphylomas were also not detected by WF-OCT images in 23 (66%) eyes (Fig. 6). In the 12 (34%) eyes in which staphylomas were detected by WF-OCT (Fig. 7), but not by 3D-MRI, the discrepancy is likely explained by the fact that the change in the curvature of the sclera was subtle so that it was missed on 3D-MRI but not on the WF-OCT images with their higher spatial resolution (Fig. 7). Of the 25 eyes for which WF-OCT did not show any staphylomas, 23 eyes did not show any staphylomas on 3D-MRI (Table). For the remaining two eyes, 3D-MRI showed wide staphylomas that were larger than the 16-mm OCT scan line so that the edge of the staphyloma was not visualized on the WF-OCT images. These two eyes were the only ones for which the staphylomas did not fit within the scan length of the WF-OCT. Both examiners agreed on the presence and type for all staphylomas detected in the study.

Comparing the classification of the staphylomas showed a congruency of both techniques for 71 eyes (71%) (Table). The concordance between both instruments was considered to be good, with a concordance index kappa of 0.61 (95% confidence interval: 0.50–0.72). Among the 75 eyes that were diagnosed as having staphylomas by WF-OCT, 29 eyes (38.7%) had wide macular staphylomas, 27 eyes (36%) had narrow macular staphylomas, 6 eyes (8%) had peripapillary staphylomas, 2 eyes (3%) had inferior staphylomas, and 11 eyes (15%) had other staphyloma types. On 3D-MRI, among the 65 eyes that were diagnosed as having staphylomas, 42 eyes (56%) had wide macular staphylomas, 17 eyes (23%) had narrow macular staphylomas, 4 eyes (5%) had peripapillary staphylomas, 1 eye (1%) had an inferior staphyloma, and 1 eye (1%) had another staphyloma type. None of the eyes included in the study had nasal staphylomas either on the WF-OCT images or on 3D-MRI.

Of the 42 eyes that were diagnosed as wide macular type by 3D-MRI, 16 eyes (38%) were characterized differently on WF-OCT. Two eyes were diagnosed having no staphylomas, four eyes with narrow macular staphylomas, two eyes with inferior staphylomas, and 22 eyes with other staphyloma types. The eyes that were diagnosed by 3D-MRI with narrow macular staphylomas or peripapillary staphylomas had a complete concordance with the assessment using the WF-OCT images. WF-OCT as compared with 3D-MRI showed more eyes with “other staphyloma types,” which were difficult to be classified into one of the five staphyloma types by WF-OCT (Table). All of these 11 eyes had a combination of peripapillary staphyloma and other types of staphyloma (10 eyes with the narrow macula staphyloma and 1 eye with an inferior staphyloma). In eyes with the combined type of a peripapillary and narrow macula staphyloma, two staphylomas were divided by a vertical ridge, which was detectable on the fundus photographs and on the WF-OCT images (Fig. 5). The combined type of a peripapillary and a narrow macula staphyloma appeared to be similar to Curtin’s staphyloma type IX; however, in 8 of the 10 eyes, the vertical ridge separating the macular staphyloma type from the peripapillary staphyloma type extended into the midperiphery of the fundus (Fig. 8). Due to spatial limitations of WF-OCT, the scleral thickness or the integrity of Bruch’s membrane could not be assessed in the present study.
DISCUSSION

In our study on eyes with pathologic myopia, WF-OCT with a scan region of 16 × 14 mm and a depth of 5 mm revealed the morphology of posterior staphylomas in highly myopic eyes. Except for two eyes, the staphylomas were visible by WF-OCT in their full extent. When the detectability of posterior staphylomas by WF-OCT and by 3D-MRI was compared, WF-OCT was superior to 3D-MRI in all but two eyes in which the width of the staphyloma was longer than the scan length of the WF-OCT. In general, both techniques allowed the shape-based differentiation of the staphylomas in a similar manner. In addition to cost, one of the major advantages of WF-OCT over 3D-MRI was the ability to visualize the tissues in a markedly higher resolution and allowed the differentiation among vitreous, retina, choroid, and sclera. In addition, 3D-MRI showed only the outer surface of the vitreous cavity because, using T2-weighted images, it visualized the shape of intraocular fluid. In contrast, WF-OCT allowed visualization of the structures of the ocular wall. In eyes with an abnormal retinal surface, such as in myopic retinoschisis, 3D-MRI, in contrast to WF-OCT, may thus not validly show the contour of the sclera.

Morphologic features of posterior staphyloma as examined by WF-OCT included a gradual thinning of the choroid from the periphery toward the edge of the staphyloma and a gradual rethickening of the choroid in direction toward the posterior pole, as well as a gradual thickening and inward protrusion of the sclera at the staphyloma edge. Thinning of subfoveal choroid is a well-known feature of myopic eyes in general22,23; however, such gradual thinning of the choroid from the periphery toward the staphyloma edge as shown in the present study has not been previously reported. It was not previously possible to visualize the choroid and sclera in a wide range of the posterior ocular segment. The findings obtained in the present study agree with the observations made on smaller posterior staphylomas (such as peripapillary staphyloma14 or inferior staphyloma due to tilted disc syndrome24) in previous studies in which conventional OCT devices with a shorter scan line length were used. These investigations suggested that the OCT features of a gradual scleral thickening and a gradual choroidal thinning at the staphyloma edge might be a consistent and useful marker to detect the edge of any type of staphylomas.

Because our study had a cross-sectional design, it has been unclear whether the morphologic features at the staphyloma edge occurred before staphyloma formation or whether they
were the consequences of the development of a staphyloma. It also remains unclear whether the choroid, sclera, or potentially another tissue (such as Bruch’s membrane) is primarily affected and responsible for subsequent staphyloma formation. In addition to choroid and sclera, it has recently been discussed that Bruch’s membrane may be an important structure leading to axial elongation, caused by a growth of Bruch’s membrane in the midperipheral region and pushing the posterior Bruch’s membrane backward. It would lead to a compression of the macular choroid and a passive elongation and thinning of the sclera, most marked at the posterior pole. The integrity of Bruch’s membrane at the staphyloma edge needs to be addressed in future studies by using a device with better resolution. Future studies may quantify the dimensions of the posterior staphylomas measuring their minimal and maximal diameters, their depth, and their shape and location in the spatial relationship to landmarks such as the optic nerve head and the macula, to provide data for biomechanical calculations and models describing the development of pathologic myopia. The formation of a staphyloma includes visually important tissues, such as the optic nerve head and the macular retina, which can significantly affect the visual prognosis of highly myopic patients. The quantitative assessment of staphylomas could thus be a step to establish treatments targeting the development of staphylomas before blinding complications occur. In addition, the relationship between a staphyloma and other tissues that could influence the scleral shape (such as vitreous, ...) was not addressed.

Table. Correlation of Types of Posterior Staphyloma Identified by 3D-MRI and WF-OCT

<table>
<thead>
<tr>
<th>OCT, 3D-MRI</th>
<th>None</th>
<th>Wide Macular</th>
<th>Narrow Macular</th>
<th>Peripapillary</th>
<th>Nasal</th>
<th>Inferior</th>
<th>Others</th>
<th>Total</th>
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<tr>
<td>None</td>
<td>23</td>
<td>2</td>
<td>6</td>
<td>2</td>
<td>0</td>
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<td>2</td>
<td>35</td>
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<tr>
<td>Wide macular</td>
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<td>26</td>
<td>4</td>
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<td>0</td>
<td>2</td>
<td>8</td>
<td>42</td>
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<tr>
<td>Narrow macular</td>
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<td>17</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>17</td>
<td>42</td>
</tr>
<tr>
<td>Peripapillary</td>
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<td>0</td>
<td>4</td>
<td>0</td>
<td>0</td>
<td>4</td>
<td>4</td>
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<tr>
<td>Nasal</td>
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<td>0</td>
<td>0</td>
<td>0</td>
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<td>0</td>
</tr>
<tr>
<td>Inferior</td>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
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<td>6</td>
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<td>2</td>
<td>11</td>
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</table>

Number of observed agreements: 71 (71.0% of the observations). Number of agreements expected by chance: 25.9 (25.89% of the observations). Kappa = 0.609. SE of kappa = 0.058. 95% confidence interval: 0.496–0.722. The strength of agreement is considered to be “good.”
intrascleral/episcleral vessels) may be an interesting theme to be examined.

When discussing the findings obtained in our study, its limitations may be taken into account. First, this study examined highly myopic patients who visited a third referral center. The results may therefore not represent the general population of highly myopic patients. Second, a quantitative histomorphometric analyses of the dimensions and shape of the choroid and the sclera were not performed, so that the findings were described in a qualitative manner. Third, due to the technical limitations of the OCT method, the reconstructed 3D-OCT images might have been imprecise in the periphery of the fundus. Fourth, despite the increased width and depth of the OCT images using the new WF-OCT technology, very large staphylomas could not be visualized by WF-OCT. An even longer scan line may be necessary to visualize all staphylomas regardless of their size and location. Future studies applying a new prototype of WF-OCT with a 20-mm scan length may address that limitation of the current investigation.

In conclusion, WF-OCT provided images of posterior staphylomas in highly myopic eyes in a resolution and wide field of view previously unachievable. WF-OCT may replace 3D-MRI in assessing posterior staphylomas, which are a hallmark of pathologic myopia.

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