Subretinal neovascularisation in eyes with localised inferior posterior staphylomas

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SUMMARY We report seven cases of subretinal neovascularisation with inferior posterior staphyloma. The upper border of the shallow staphyloma, detected by B-mode echography, invaded the macular area, and there was a neovascular net at the edge of the staphyloma. Three of the cases showed chorioretinal atrophy at the upper edge of the staphyloma in the same location as the neovascular net. Our cases were identical to those of subretinal neovascularisation in high myopic eyes in which the rupture of Bruch’s membrane was related to the deformity of the scleral shell. As our cases included not-high-myopic eyes, the results suggest that the identical mechanism might be involved in the occurrence of neovascular maculopathy in the pathological high myopic eye and in the not-high-myopic eye with posterior staphyloma. Localised inferior posterior staphyloma may be an important cause of ‘idiopathic’ subretinal neovascularisation.

Subretinal neovascularisation is triggered by the rupture of Bruch’s membrane in the posterior fundus. High myopia has been described as a cause of subretinal neovascularisation, as have aging, trauma, uveitis, angioid streaks, and macular dystrophies. In the high myopic eye the uvea and retina at the posterior fundus expand so that the axial length of the eye is elongated. This condition leads to the rupture of Bruch’s membrane and the growth of subretinal neovascularisation. However, when a localised posterior staphyloma exists, the posterior fundus may expand somewhat without significant elongation of the axial length. For example, the tilted disc syndrome is not necessarily associated with high myopia but has the typical high myopic fundus in the lower half of the eye related to the inferior staphyloma. In such a case myopic pathological complications may occur inside the posterior staphyloma even if the eye is not high myopic.

We report here a series of consecutive cases with subretinal neovascularisation that are not severely high myopic but have inferior posterior staphylomas in order to study the relationship between the inferior staphyloma and subretinal neovascularisation.

Material and methods

We examined 32 patients with subretinal neovascularisation of unknown origin between 1980 and 1982 and found that seven had inferior posterior staphylomas. High myopic cases of −8.25 D or more were excluded. Indirect binocular ophthalmoscopy and B-mode echography were used to detect the staphyloma. The ultrasound equipment was Coleman Ophthalmoscan Model 200 (Sonometrics). We used a 10 MHz focused transducer for B-mode echography. Axial length measurement was performed by DBR-300 (Sonometrics), where the velocity of sound in the eye was decided to be 1550 m/s.

To estimate the existence and extent of subretinal neovascularisation we performed fluorescein angiography. Refractive power was measured by an ultrared autorefractometer (TOPCON). Serological tests for toxoplasmosis, leptospirosis, and syphilis were performed.

Results

A profile of the seven patients is shown in Table 1. Serological tests for toxoplasmosis, leptospirosis, and syphilis were negative, and no active or atrophic focus of inflammation, such as the punched-out lesions found in presumed ocular histoplasmosis
syndrome, was detected by fundus examination. Other diseases that may cause subretinal neovascularisation such as trauma, angiod streaks, and macular dystrophies were not found in these cases. Refraction ranged from +1.25 D to −8.0 D. Five of seven cases had vertical astigmatism. Refractive power within the inferior posterior staphyloma was also measured, so that all cases were found to be more myopic inferiorly.

The fibrovascular membranes were small, ranging from 0.3 disc diameter to 1.0 disc diameter. Subretinal or choroidal haemorrhage around the fibrovascular membrane was seen in four cases (cases 1, 2, 3, 7). A chorioretinal atrophic focus lying across the macular area horizontally was seen in three cases (cases 1, 3, 6). The atrophic focus formed the boundary between the upper fundus, with normal colour, and the lower one, with a tigroid appearance. Crescent formations were seen in all cases.

The posterior staphyloma detected by B-mode echography was localised in inferior half of the posterior pole and was most clearly shown when the eye was sectioned vertically. Representative echograms are shown in Figs. 1 and 2. Staphylomas were shallow and small in size. The upper edge of the staphyloma was sharp, lying across the macular area.

Representative fluorescein angiograms are shown in Figs. 3 and 4. Subretinal neovascularisation was seen as typical fibrovascular nets at the macular area in the early phase of fluorescein angiography. We could also detect the inferior posterior staphyloma in fluorescein angiography with the aid of its sharp focusing. As the depth of the fundus inside the staphyloma was different from that outside it, the sharpness was guaranteed in half of the photograph, either inside or outside the staphyloma. Fluorescein angiography showed that fibrovascular membranes were close to the upper edge of the inferior posterior staphylomas in all cases. Atrophic foci seen in three cases were round the sharp upper edge of the staphyloma, and showed hyperfluorescence of a window defect (Fig. 3).

Unaffected eyes also had inferior posterior staphylomas (seven eyes), vertical astigmatism (five eyes), and inferior or temporal inferior crescents (seven eyes).

Discussion

With regard to the subretinal neovascularisation in high myopia Curtin and Karlin reported that the regression coefficient for the incidence of Fuchs’s spot in the eyes measuring 25-5 to 36-6 mm axial length showed no statistical significance. More recently Hotchkiss and Fine noted that the median refractive error in a group with subretinal neovascularisation was less severe than in a group of unaffected eyes of identical cases. We reported that among myopic macular complications subretinal neovascularisation was seen when the posterior pole was not too elongated compared with macular break or focal chorioretinal atrophy. These findings suggest that subretinal neovascularisation occurs when the posterior pole of the eye is moderately elongated and that it is not necessarily high myopic. Hence, even if our seven cases include a hyperopic

Table 1 Profile of seven cases

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>Eye</th>
<th>Visual acuity</th>
<th>Refraction (D)</th>
<th>Axial length (mm)</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Vertical</td>
<td>Horizontal</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>56</td>
<td>F</td>
<td>L</td>
<td>16/200</td>
<td>+0.25</td>
<td>+1.25</td>
<td>23-6</td>
</tr>
<tr>
<td>2</td>
<td>33</td>
<td>F</td>
<td>R</td>
<td>100/200</td>
<td>−0.5</td>
<td>−5.0</td>
<td>25-8</td>
</tr>
<tr>
<td>3</td>
<td>54</td>
<td>M</td>
<td>L</td>
<td>80/200</td>
<td>−0.2</td>
<td>−1.5</td>
<td>23-8</td>
</tr>
<tr>
<td>4</td>
<td>46</td>
<td>F</td>
<td>L</td>
<td>60/200</td>
<td>−0.5</td>
<td>−5.0</td>
<td>26-0</td>
</tr>
<tr>
<td>5</td>
<td>28</td>
<td>F</td>
<td>R</td>
<td>16/200</td>
<td>−0.5</td>
<td>−5.0</td>
<td>25-0</td>
</tr>
<tr>
<td>6</td>
<td>59</td>
<td>F</td>
<td>R</td>
<td>2/200</td>
<td>−0.5</td>
<td>−2.5</td>
<td>24-0</td>
</tr>
<tr>
<td>7</td>
<td>35</td>
<td>F</td>
<td>L</td>
<td>30/200</td>
<td>−1.5</td>
<td>−1.5</td>
<td>24-1</td>
</tr>
</tbody>
</table>

* Refraction within the staphyloma in parentheses.
† Disc diameter.
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eye, it is probable that they are identical to cases of subretinal neovascularisation in high myopia.

Three cases with atrophy at the edge of the staphyloma have the same characteristics, including relatively low refractive power, occurrence in later middle age, and a sharp upper border of the staphyloma drawn in vertically sectioned B-mode echogram. These cases demonstrate the relation between subretinal neovascularisation and inferior posterior staphyloma, for the neovascular nets are just on the atrophic focus round the upper edge of the staphyloma. A sharp curve round the border of the staphyloma may cause a localised degeneration of the choroid and retina, resulting in the breakdown of Bruch’s membrane and subretinal neovascularisation when the macular area is involved.

 Inferior posterior staphyloma, the main characteristic of our cases, is also found in the tilted disc syndrome. Cases 1 and 3 fit the tilted disc syndrome exactly, because they have the typical bilateral visual field defect. In the other five cases the existence of an inferior posterior staphyloma and inferior or temporal inferior crescent show their resemblance to the tilted disc syndrome. Why a visual field defect occurs in the tilted disc syndrome is still unclear, but, as the defect is in accordance with the inferior posterior staphyloma, the extension of the posterior fundus may cause the nerve fibre bundle defect. Similarly, Bruch’s membrane may break down inside or at the edge of the staphyloma. Tilted disc and inferior or temporal inferior crescent are reliable signs of the existence of inferior posterior staphyloma.

The large and deep staphyloma in high myopia can be seen easily by binocular ophthalmoscopy, but a shallow one is sometimes difficult to detect. Some of the shallow staphylomas in our cases could be detected
only by ultrasonic echography. In the echographic
drawing of the shape of a shallow staphyloma it is
important to know where the staphyloma is located.
We could get a clear drawing of the staphylomas by
vertically sectioned B-mode echogram, since the
staphylomas were always located at the lower half of
the posterior fundus. The direction of the staphyloma
almost coincided with that of the crescent, indicating
the location of the staphyloma.

The age distribution of our patients (from 28 to 59
years) is noteworthy. First, senile disciform macular
degeneration can be excluded. Secondly, as changes
similar to high myopia are characteristic of senile
eyes of any refractive denomination, we could not
decide whether the change was related to the
posterior staphyloma if the patient was 60 or over.

As our cases distinctly show that localised inferior
posterior staphyloma is associated with subretinal
neovascularisation, we assume that it is an important
and perhaps main cause of 'idiopathic' subretinal
neovascularisation.

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