Heterotopia of the Macula with Associated Retinal Detachment*

A Case Report
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Many cases of heterotopia or ectopia of the macula result from an abortive form of retrolental fibroplasia (ARF). The gliosis which occurs following oxygen toxicity causes a distortion of the vessels with subsequent disturbance of the normal retinal vascular anatomy and a displacement of the macula. This distortion of vessels is particularly prominent about the disc and results in a straightening of the normally tortuous vessels. This most commonly occurs on the temporal side of the retina. It gives the appearance that the optic disc and vessels have been dragged toward the periphery.

One would expect that such dragging and traction would frequently be accompanied by tears in the retina. However, to the best of the authors' knowledge, no case of heterotopia of the macula with an associated retinal detachment has been reported.

Case Report

The patient (A.K.) an 18-year-old white male, was first seen on June 17, 1967, after being referred by his own ophthalmologist. The patient related that he had not seen well from his left eye since Thanksgiving 1966. There was a history of minor trauma involving the forehead seven months earlier; there were no immediate sequelae. The only significant past history indicated that he was born prematurely and required supplemental oxygen administration after birth.

Examination revealed a corrected vision O.D. of 20/100 and O.S. of 20/200. He was wearing O.D. -7.00 -2.00 x 62 and O.S. -7.25 sphere. Lids, lashes, and lacrimal apparatus were normal, as were the corneas and anterior chamber; the lenses were clear. The pupils showed a partial mydriasis from medication. No holes were seen in the right periphery. The vessels on the disc were dragged somewhat superiorly, as was the macula.

Examination of the left retina revealed an inferior detachment involving the macula, and demarcation lines in the upper temporal retina. A horseshoe tear was noted at the 3:30 position anterior to the equator. The lower third of the retina, anterior to the equator, was devoid of visible arterioles and venules. The retinal circulation appeared to end along a line anterior to the equator with slight dilatations at the terminations of the vessels. The posterior attachment of the vitreous base could be seen posteriorly to this line. (Figs. 1 and 2). The vessels about both discs were dragged superiorly (Figs. 3 and 4), and displacement of the maculae superiorly was also evident (Figs. 5 and 6). This condition of the macula was not immediately recognized because of the detachment. (Fig. 7). Preoperative visual fields (Fig. 8) revealed an irregular defect. He showed six-prism dioplers of exophoria and four-prism dioplers of right hyperphoria at 20 ft., and 25-prism dioplers of exotropia and eight-prism dioplers of right hypertropia at 13 inches. He preferred the right eye. Near point of convergence was remote. ERG showed slight reduction O.D. and marked reduction O.S. (Table I).

On the following day, June 18, 1967, the patient was admitted to the hospital for bed rest and observation but failed to show any improvement. Two days later, he underwent a repair of his retinal detachment by encircling scleral buckle and transcleral cryocoagulation with the Kelman Retinal Unit. Drainage was performed.

Postoperatively, the hole remained open. Conse-
Fig. 1. Detachment of retina with peripheral avascularity and terminal glomeruloid tufts O.S.

Fig. 2. Detachment of retina O.S., as in Figure 1, showing the posterior attachment of the vitreous base.

Fig. 3. Right disc showing dragging of vessels superiorly.

Fig. 4. Left disc showing dragging of vessels superiorly.

Fig. 5. Macular displacement superiorly O.D.

Fig. 6. Macular displacement superiorly O.S.
Fig. 7. Preoperative detachment (schematic drawing). Gray area indicates detached retina.

Fig. 8. Preoperative visual fields.

Fig. 9. Postoperative retinal detachment (schematic drawing).

Table I

<table>
<thead>
<tr>
<th>ELECTRORETINOGNOSY</th>
<th>O.D.</th>
<th>O.S.</th>
<th>Normal Range</th>
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<tr>
<td>A wave</td>
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<td>20</td>
<td>87 - 149</td>
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<tr>
<td>B wave</td>
<td>138</td>
<td>39</td>
<td>164 - 284</td>
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<td>0</td>
<td>39 - 77</td>
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<td>D wave</td>
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<td>20</td>
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<tr>
<td>Dr 16</td>
<td>53</td>
<td>0</td>
<td>51 - 97</td>
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corrected, was 20/50-1 and 20/100 O.S. Pin-hole vision O.S. was 20/60-2. He showed approximately seven degrees of hypertropia by corneal reflex. He measured five-prism diopters of right hyperphoria at 13 inches, and three- to four-prism diopters of right hyperphoria by screen and cover test. Troposcope measurements showed third degree fusion with six-prism diopters of convergence and 14-prism diopters of divergence.

Discussion

Heterotopia of the macula is an unusual finding although not quite as rare as once thought. Any significant alteration in the anatomic location of the macula in relation to the optic disc or variation in angle Kappa is considered heterotopia of the macula. Various etiologies have been suggested, and Rados and Scholz1 did an extensive review of the existing world literature. They tabulated 27 acceptable
cases, including eight of their own. Their summary of the 27 cases showed that only ten cases were bilateral heterotopia of the macula. Their analysis revealed that three cases were congenital in origin; two, inflammatory; ten, chorioretinitis; and twelve were probably due to incomplete or an abortive form of retrolental fibroplasia (ARF). None was associated with a retinal detachment.

The relationship of heterotopia of the macula to abortive retrolental fibroplasia (ARF) was first described by Payne and Crick. Six more cases of heterotopia were described by Nauheim, four of which were consistent with an etiology of prematurity and incubation in oxygen, resulting in ARF. Another similar case was also considered the result of prematurity. Since the report by Nauheim, several other cases have been reported. A recent article by Backus and included a searching review and classification of heterotopia of the macula.

Rados and Scholz listed seven useful points to aid in making the clinical diagnosis of heterotopia of the macula:

1. On fixation with both eyes, the eye with the ectopic macula will tend to deviate in the same direction as the macular displacement.
2. Central fields will show displacement of the blind spot corresponding to altered anatomic relations between the disc and the macula.
3. Cover-Uncover Test will show no shift, or a strabismus opposite that which was thought to exist (pseudostrabismus).
4. Although there appears to be eccentric fixation, the patient will fix with his macula.
5. On fundus examination, there is an abnormal location of the macula in relation to the optic disc.
6. Visual acuity will be from normal to low.
7. When an actual strabismus exists and the macula is not damaged, an amblyopic eye may respond to patching.

An interesting feature of this case was the zone of peripheral avascularity; this was probably the result of an obliteration of the terminal arterioles prior to their reaching the retinal periphery during development. This obliteration was associated with a high oxygen concentration in the retina. Glomeruloid capillary tufts were seen at the termination of the peripheral vessels.

Factors which may have contributed to the retinal detachment were myopia, the avascular peripheral retina, and vitreous traction resulting in the horseshoe tear. No explanation for the ERG changes can be given at this time. The macula was displaced superiorly more than temporally. This resulted in a pseudostrabismus simulating a left hypertropia of seven degrees as evidenced by the corneal reflex. The heterotopia of the right macula was slight, and no obvious pseudostrabismus could be measured.

In this patient, however, the authors felt that the history of prematurity and supplemental oxygen therapy, plus the clinical appearance, were consistent with a diagnosis of incomplete or abortive retrolental fibroplasia, producing a heterotopia of both maculae. The history of minor trauma to the head may have precipitated the detachment of the retina, which had already undergone traction from retrolental fibroplasia (RLF).

It is known that RLF can cause retinal breaks and detachments, as well as heterotopia of the macula. These two manifestations of RLF could logically result from the same process: stretching of the retina. It is surprising, therefore, that this is the only case in which the two effects coincide in the same eye.

Summary

An 18-year-old boy, born prematurely and incubated in an unknown supplemental oxygen concentration, developed an abortive form of retrolental fibroplasia with resultant bilateral heterotopia of the maculae and an associated retinal detachment. A minor head trauma, seven months previously, may also have been a precipitating factor in the pathogenesis of the retinal detachment. To the best of the authors' knowledge, no previously published case of heterotopia of the macula has been reported with a coinciding retinal detachment in the same eye.

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References


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