vascular masses occurring in the peripheral retina have been described extensively in the literature. Many terms, including “presumed acquired hemangiomas,” “hemangioma-like,” “angiomatous masses,” “angioma-like,” “peripheral retinal telangiectasis,” and “vasoproliferative tumors,” have been suggested that reflect the lack of the known histological features and the potentially variable causes. We describe the histological features of 2 patients who underwent transcleral local resection as management for suspected choroidal melanoma. Pathological examination of these tumors reveals the constituents to be primarily benign glial cell proliferation with secondary vasoproliferation. The weight of the literature agrees with a reactionary process. We therefore suggest the term “reactionary retinal glioangiosis.” Transcleral resection has a place where diagnosis is difficult. It prevents an unnecessary enucleation and allows accurate tissue diagnosis.

In 1983 Shields et al1 coined the term “presumed acquired retinal hemangiomas” to describe solitary peripheral retinal vascular masses that resembled either retinal hemangiomas or choroidal melanomas. The initial study was in a small group of patients, but a later review of a larger group of 103 patients by Shields et al2 led to a change in nomenclature, with the authors renaming these lesions “vasoproliferative retinal tumors.” They proposed a classification based entirely on clinical features. These studies were not supported by histopathological features although Welch,3 in discussion of the 1983 paper by Shields et al,1 suggested that these tumors might be similar to lesions described in 1966 by Henkind and Morgan4 in 4 enucleated eyes.

Since morphological descriptions of this clinical entity are therefore sparse, we describe the pathological features of 2 tumors that conform to the clinical descriptions provided by Shields and coworkers.1,2 The patients were referred to the Scottish National Ocular Oncology Service, Glasgow, for the treatment of suspected malignant melanomas of the peripheral choroid. The differential diagnosis of these cases included vasoproliferative retinal tumors; however, since the diagnosis of malignant melanoma could not be excluded, the tumors were removed by transcleral resection. In this article we describe the clinical and histopathological features of these rare benign abnormalities.

REPORT OF CASES

CASE 1

A 61-year-old man was referred in September 1997 with the diagnosis of a presumed choroidal melanoma in the inferotemporal quadrant of the right eye. Five months previously he had noticed distortion and decreased vision in this eye. On ophthalmic examination visual acuity was reduced to counting fingers OD and 20/20 (6/6) OS. There was a right afferent pupil defect. Funduscopy revealed a solid-looking lesion in the inferotemporal quadrant of the right eye with associated exudative retinal detachment and retinal hemorrhage (Figure 1). No obvious feeder or draining vessels were noted. A definite transillumination defect associated with the lesion was noted. Fundus fluorescein angiography showed late hyperfluorescence only. Ultrasound B-scan...
demonstrated a 6.3 × 5.6-mm choroidal mass that showed moderate to high internal reflectivity with associated retinal detachment; the images were consistent with a choroidal melanoma (Figure 2). Results of systemic investigations were normal; there was no family history of ocular disease. The patient was not hypertensive.

Although the clinical appearance was not entirely typical of a choroidal melanoma, this diagnosis could not be excluded and a decision was made to locally resect the mass and to apply a radioactive ruthenium 106 plaque over the resection site.

Preoperatively the tumor did not involve the choroid as evidenced by the lack of adhesion between the scleral trapdoor and an apparently normal choroid. The diagnosis of choroidal melanoma was therefore immediately in doubt. The tumor was excised as planned because the aim was to preserve vision and the procedure was completed by the technique described below in the “Transcleral Resection” subsection of the “Materials and Method” section.

Because the initial histological examination revealed a vasoproliferative tumor with no evidence of malignancy, the ruthenium 106 plaque was removed 2 days postoperatively. Since similar lesions are found in von Hippel-Lindau syndrome, appropriate investigations were carried out and the results were normal.

At follow-up assessment 4 months postoperatively, although visual acuity was restricted to hand movements, subjectively the patient felt there had been a noticeable improvement. The retina had anatomically flattened, the vitreous was much clearer, and a marked resolution of the retinal vascular reaction was noted (Figure 3). The patient remains stable at 18 months' follow-up.

CASE 2

A 63-year-old man was referred in November 1998 with a right inferonasal raised mass presumed to be a choroidal melanoma. He had noticed gradual reduction of vision in this eye. On ophthalmic examination visual acuity was 20/130 (6/36) OD and 20/20 (6/9) OS. Funduscopy revealed a yellow crystalline inferonasal mass with associated exudative retinopathy (Figure 4). No obvious feeder or draining vessels were noted. Ultrasound B-scan confirmed a solid lesion 10 mm across the base and 5 mm in depth with associated vitreous debris consistent with a choroidal melanoma (Figure 5). Fluorescein angiography was not performed. Results of systemic investigations were normal; there was no family history of ocular disease.

Local resection surgery with brachytherapy was planned and carried out as described below in the “Transcleral Resection” subsection of the “Materials and Methods” section. Again, preoperatively no choroidal or scleral involvement was found, putting the initial diagnosis in question. The surgical procedure was completed without complication. The ruthenium 106 plaque
was removed 3 days postoperatively once the pathological diagnosis was provided and confirmed that there was no evidence of malignancy.

At the 1-month follow-up visual acuity remains 20/130 (6/36) OD and the retina remains flat (Figure 6). The patient remains stable at 6 months’ follow-up.

MATERIALS AND METHODS

TRANSCLERAL RESECTION

The surgical approach of transcleral local resection involves localization of the tumor by indirect ophthalmoscopy followed by formation of a lamellar scleral flap. The tumor is then dissected from the retina and excised along with the deep scleral lamella and an area of normal surrounding choroid, avoiding retinal damage. The scleral flap is replaced and sutured in position.

Intravitreal sulphurhexafluoride is injected to provide retinal tamponade. Hypotensive anesthesia with the systolic blood pressure being lowered to around 50 mm Hg is obligatory.5,6

PATHOLOGICAL FEATURES

The excised retinal mass, the attached choroid, and the inner scleral flap in both cases were fixed in buffered 2.5% glutaraldehyde and processed for paraffin histological examination.

Paraffin sections were stained with a variety of routine stains including hematoxylin-eosin, periodic acid–Schiff, trichromes (Mallory, Masson, or picro-Mallory), reticulin, Martius scarlet blue, and the elastica van Gieson. Immunohistochemical markers were used to identify the glial component (glial fibrillary acidic protein, S100 protein, and neurofilaments) and the endothelial cells (QB10) in the blood vessels within the tumor.

RESULTS

CASE 1

Macroscopically the mass measured 7 × 4 × 3 mm and consisted of soft-folded, pale brown hemorrhagic tissue.

Microscopic examination demonstrated that the tumor consisted of a large mass of spindle cells surrounding blood vessels of various sizes (Figure 7, A-D). The spindle cells were of uniform size and mitotic figures were absent: positive staining with phosphotungstic hematoxylin and glial fibrillary acid protein confirmed the glial nature of these cells (Figure 7, D). In
A large mass of fibrin and granulation tissue was identified (Figure 7, B). The major part of the vascular component took the form of clusters of channels lined by plump endothelial cells, within hyalinized connective tissue (Figure 7, B and D). Small angiomatoid capillary clusters were also present (Figure 7, C) within gliotic areas and at the periphery of the mass. Fibrinous, lipid, and serous exudates were present in the tissue adjacent to the blood vessels. Extensive areas of “basement membrane deposit”7 (synonyms: basal linear deposit, basal lamina deposit) were present on the inner surface of Bruch’s membrane (Figure 7, E). Elsewhere Bruch’s membrane was lined by a fibroctic membrane due to proliferation and metaplastic change within the retinal pigment epithelium (RPE) (Figure 7, F). A few foci of neovascularization were noted in this region (Figure 7, F). Retinal tissue containing hemorrhage and exudates was identified at 1 periphery. Abnormal tissue extended to the edge of the mass in most paraffin blocks.

CASE 2

Macroscopically a tan lobulated mass (Figure 8, A) was attached to a flatter region in which there were pale yellow crystalline areas. The total dimensions of the mass were 6 × 5 × 3 mm. Blood vessels were prominent on the surface of the mass.

On microscopic examination the main mass of the tumor was formed by spindle cells in an irregular arrangement (Figure 8, B-D). Cystic spaces surrounded by macrophages were identified in the flatter region; these corresponded to the yellow loci shown in Figure 8, A.

Blood vessels within the tumor were frequently associated with a perivascular lymphocytic infiltration and this was associated with the high endothelial venule phenomenon. Examples of thrombosis and intraluminal endothelial cell proliferation and fibrin deposition were identified; many blood vessels were hyalinized and deposits of fibrin were plentiful in the inner part of the tumor (Figure 8, C and E). A feature of interest was a focus of RPE proliferation within the glial mass (Figure 8, F). These appearances were consistent with a peripheral retinal vascular tumor. In this case the tumor tissue extended to the ex continued.
similar to vascular tumors, as described by Shields et al. 17

Pathological examination of the 2 excised specimens also revealed the constituents to be blood vessels that varied in size from large venules to smaller vessels surrounded by thick connective tissue sheaths. Some of the vessels contained fibrin thrombi with early endothelial cell proliferation and organization. In both cases the vessels were surrounded by lymphocytes and the endothelial cells showed the high endothelial venule change. A few angiomatoid clusters were also observed. The glial cells within the mass appeared to be of a completely benign nature.

The presence of basement membrane deposit at the retinal periphery in patient 1 but not in
Patient 2 is a frequently recognized histological abnormality and is presumably irrelevant to the retinal changes. It may be implicated in peripheral subretinal neovascularization that has been described in a wide spectrum of ocular disease.

The histological features of these cases have allowed a more specific interpretation of these peripheral retinal vascular lesions. We suggest that the new term—reactionary retinal glioangiosis—would seem a more appropriate descriptive term for the clinical entity previously classified as primary vasoproliferative retinal tumor. This supports the
CONCLUSION

Both of our patients underwent surgery as management for suspected malignant melanomas. It was clear during the surgical procedure that the diagnosis was in question because of the absence of choroidal involvement. However, in cases in which the presence of a malignant melanoma cannot be excluded, local transcleral resection can avoid an unnecessary enucleation and also provides tissue to enable definitive histological diagnosis. We believe such a procedure has a place in the management of these peripheral vasoproliferative retinal tumors that form an important part of the differential diagnosis of an intraocular mass.

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REFERENCES


Ophthalmological Numismatics

A look at the past...

Albrecht Mooren, 1828-1899, was head of the Municipal Eye Hospital, Dusseldorf, Germany, from 1862 to 1883. He also became director of the Ophthalmic Institute of Liege from 1868 to 1878. The medal shown was engraved by Charles Weiner in 1880. The obverse (Figure 1) depicts the head of Morren facing left; the reverse (Figure 2), the god of light, a winged youth flying to the right, with a star above his head and a floating ribbon around his neck. In his left hand, he holds a radiant sun; in his right, a head bandage. The Latin inscription, E Tenebris ad Lucem (which translates to “Out of darkness to light”) surrounds the reverse. The obverse portrait of this medal was used as a model for the monument to Mooren that was later erected by the city of Dusseldorf.

Courtesy of: Jay M. Galst, MD, 30 E 60th St, New York, NY 10022.