Neoplasms of the Retinal Pigment Epithelium

The 1998 Albert Ruedemann, Sr, Memorial Lecture, Part 2

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Background: Neoplasms of the retinal pigment epithelium (RPE) are rare, and little is known about their clinical variations, clinical course, and prognosis. Although most are benign, they can simulate choroidal melanoma.

Objectives: To evaluate the clinical characteristics, management, pathological features, and prognosis of acquired neoplasms of the RPE (adenoma and adenocarcinoma) and to define clinical features that help differentiate them from uveal melanoma.

Patients and Methods: The medical records of patients with acquired tumor of the RPE were reviewed retrospectively, and the clinical features, management, and histopathologic findings were studied.

Results: Of 13 affected patients, 10 were women and 3 were men. Ten were white and 3 were African American. The mean age at diagnosis was 53 years (range, 28-79 years). All patients were referred with the diagnosis of suspected choroidal melanoma. All tumors were solitary, unilateral, and ranged from a small, asymptomatic lesion measuring 2 x 2 x 1 mm to a massive neoplasm that measured 17 x 17 x 17 mm. There was no predilection for retinal location or laterality. The tumors were dark brown to black in 11 patients and only minimally pigmented in 2. Prominent retinal feeder vessels were visualized in 8 patients, 5 of whom had an exudative retinal detachment. Two patients had recurrent vitreous hemorrhage. Transillumination generally revealed blockage of light by the tumor. Fluorescein angiography showed early hypofluorescence and late minimal hyperfluorescence of the tumor, without visibility of choroidal vessels. Ultrasonography typically demonstrated the tumor to be abruptly elevated and to have medium to high internal reflectivity and acoustic solidity. Results of diagnostic fine needle aspiration biopsy, performed on 4 patients, disclosed cells compatible with a pigment epithelial origin. Treatment ultimately included observation in 4 patients, enucleation in 3, local tumor resection in 3, irradiation in 2, and laser therapy in 1. Microscopic verification of the diagnosis was available in 3 eyes after fine needle aspiration biopsy, 3 eyes after local resection, 3 eyes after enucleation, and 1 eye post mortem. The microscopic diagnosis was adenoma in 8 patients and adenocarcinoma in 2. Microscopically, the lesions were composed of a neoplastic proliferation of RPE cells. Tumors arising from the anterior portion of the RPE had a vacuolated pattern, and those in the posterior portion of the RPE had a glandular or tubular configuration.

Conclusions: Neoplasms of the RPE show considerable clinical variation. In contrast to melanoma, they generally are darker, more abruptly elevated, and more likely to have retinal feeder vessels and exudative retinal detachment; show early hypofluorescence and mild late hyperfluorescence on angiographic findings; and have high internal reflectivity on ultrasonographic findings. Although most acquired tumors of the RPE are benign cytologically, they can exhibit aggressive clinical behavior.


RESULTS

The pigment epithelium of the iris (IPE), ciliary body (CPE), and retina (RPE) can undergo reactive hyperplasia, but true neoplasms of the pigment epithelium are rare. The clinical and pathological features of tumors of the IPE and CPE were recently reported.

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With the exception of 1 pathological series, most reports of tumors of the RPE have been case descriptions or very small series. We report herein our personal experience with 13 neoplasms (adenoma and adenocarcinoma) of the RPE, describe new observations of their clinical variations, and elaborate on the features that help differentiate neoplasms of the RPE from choroidal melanoma and other pigmented fundus lesions.

There were 13 patients in our files with the diagnosis of adenoma or adenocarcinoma of the RPE (Table 1). The mean age at diagnosis was 53 years (range, 28-79 years). Three patients were men and 10 were women; 10 were white and 3 were African American.

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PATIENTS AND METHODS

The medical records of our patients with the diagnosis of adenoma or adenocarcinoma of the RPE were retrieved, and the clinical features, course, and management were reviewed. We excluded cases of adenoma of the CPE, congenital hypertrophy of the RPE, and combined hamartoma. General data collected included patient age, sex, race, referring diagnosis, previous management, history of ocular trauma or inflammation, associated ocular disease, associated systemic disease, ocular symptoms, and visual acuity. Three of the 13 patients were previously described in individual case reports. We categorized the location of the epicenter of the tumor as juxtapapillary (touching the optic disc), macular (within the temporal retinal vascular arcades, midperipheral (between the arcades and equator), or peripheral (between the equator and ora serrata). Other tumor data included size ( basal diameter and thickness in millimeters), color, side effects on adjacent structures, transillumination features, fluorescence ophthalmoscopic findings, and ultrasonographic characteristics.

Histopathologic features reviewed included results of fine needle aspiration biopsy (FNAB) (when performed) and results of histopathologic studies (when available). We reviewed available histopathologic preparations and categorized the tumors as solid, vacuolated, or combined types. We determined the degree of pigmentary, vascularity, nuclear atypia, mitotic activity, and tumor invasiveness. The method of management and results were determined. Based on these observations, we make recommendations regarding the clinical diagnosis and management of acquired neoplasms of the RPE.

Other findings in patients with RPE tumors are shown in Table 1. A prominent retinal artery and retinal vein supplied and drained the tumor in 8 patients (Figure 1, E-H). These vessels were clearly visualized on fluorescein angiography (Figure 2). The patients in whom such vessels did not show included the 3 with juxtapapillary tumors (patients 1, 6, and 13) and the 2 in whom the tumor was not clearly visualized ophthalmoscopically (patients 8 and 11). Overall, prominent tumor-associated retinal vessels were present in 8 peripheral or midperipheral tumors. All 3 patients who had the most prominent, dilated, tortuous vessels were African American (patients 4, 5, and 7).

An exudative retinal detachment was observed in 5 patients. Two of these (patients 5 and 7) had total retinal detachments that did not resolve after plaque radiotherapy for the tumor. Only patient 8 had a secondary cataract.

Other findings in eyes with tumors of the RPE are listed in Table 1. Nonspecific choroiditis was found in patient 2 and inactive histoplasmic choroiditis in patient 6. Other findings included vitreous hemorrhage and fibrosis (patients 2 and 4), surface wrinkling retinopathy (patient 4), and macular hole (patient 10).

Transillumination showed blockage of light by the tumors. Fluorescein angiography, performed in 9 patients, typically showed early hypofluorescence of the tumor, with continued relative hypofluorescence and leakage into the vitreous in the late frames (Figure 2). None of the tumors showed prominent choroidal vessels (double circulation) that typify some melanomas. Ultrasonography, performed in 11 patients, demonstrated high to medium internal reflectivity with A-scan and acoustic solidity and a dome-shaped configuration with B-scan (Figure 3). Results of FNAB, performed in 4 patients, demonstrated cells compatible with a tumor of the RPE in each instance.

The tumors of the RPE showed characteristic, but variable, pathological features (Figure 4). Grossly, they were usually deeply pigmented and abruptly elevated (Figure 4, A-C). Histopathologic or cytopathologic verification of the diagnosis was available in 10 patients (Table 1). Histopathologically, all lesions appeared to arise abruptly from the adjacent normal RPE (Figure 4, D and E). Of the 7 entire tumors available for microscopic study, 3 had a solid pattern, 2 had a vacuolated pattern, and 2 had a combined pattern. The vacuolated tumors (Figure 4, D-F) were located in the more anterior region of the RPE, and the solid tumors (Figure 4, G and H) were more posteriorly located (Table 2). The vacuolated tumors had numerous round, clear vacuoles that contained a hyaluronidase-resistant acid mucopolysaccharide. The solid tumors consisted of ribbons or tubules of cells separated by connective tissue septae (Figure 4, D-H) with abundant basement membrane production that was readily observed with periodic acid–Schiff preparations. Only patient 11 had a tumor with appreciable mitotic activity.

Local invasion of the tumor into the adjacent uveal stroma, sensory retina, and vitreous was observed in 6 of the 7 tumors for which adequate sections were available. In patient 11, the tumor had extended through a staphylomatous defect into the orbit. Most tumors showed mild variation in nuclear and nucleolar size. Mitotic figures were demonstrated in patient 11.
tion between adenoma and adenocarcinoma was generally difficult. However, based on opinions by several ophthalmic pathologists, 8 tumors were ultimately classified as adenoma and 2 as adenocarcinoma, based on degree of nuclear atypia and degree of local invasiveness.

Cytologic diagnosis from results of FNAB was obtained in 4 patients, 1 of whom later underwent enucleation. In patient 6, the diagnosis of adenocarcinoma was made and later confirmed histopathologically. Overall, a total of 10 patients had microscopic verification (histopathologic or cytopathologic) of the diagnosis of tumor of the RPE. In the other 3 patients, the diagnosis was based on the classic clinical findings.

**COMMENT**

Although reactive hyperplasia of the RPE is well known, true neoplasms of the RPE are uncommon.\(^1\)\(^,\)\(^2\) Hyperplasia of the RPE can assume tumorous proportions and be difficult to differentiate clinically and histopathologically from a true neoplasm.\(^3\)\(^,\)\(^4\)\(^,\)\(^20\)

A pigmented process is likely to be a reactive hyperplasia if there are clinical or pathological signs of trauma or inflammation. If such signs are absent, then the diagnosis of true neoplasm should be considered. None of our patients had convincing ocular signs of trauma. Although 2 of our patients had signs of mild intraocular inflammation in the affected eye, the tumors in both were studied histopathologically and were interpreted by ocular pathologists as true neoplasms, rather than reactive hyperplasia. Based on our observations and a review of the literature, we believe that most tumors of the RPE occur in otherwise healthy eyes, but that they can occasionally arise at a site of previous ocular inflammation, possibly due to chronic hyperplasia of the RPE.

Our experience with these patients, combined with a review of the literature,\(^7\)\(^-\)\(^28\) has allowed us to better understand the demographics, clinical features, clinical course, management, pathological features, and prognosis of neoplasms of the RPE.

A neoplasm of the RPE is usually diagnosed in adulthood. A review of previously reported cases revealed a mean age of 49 years at the time of diagnosis. In our series, the mean age at diagnosis was 53 years (range, 28-79 years). However, we cannot exclude the possibility that some of these tumors were congenital and were not detected until adulthood, when they became sizable enough to cause ocular symptoms. Patient 1 had a tumor that was diagnosed as a melanocytoma and was observed without change for 10 years. The diagnosis of adenoma of the RPE ultimately was made after postmortem examination of the eye.

Our series is too small to determine if there is a predilection for race and sex, and the literature does not always mention the race and sex of the patients. Three of our 13 patients were African American and 10 (77%) were women. A review of acceptable cases in the literature that denoted sex disclosed 12 women and 5 men with neoplasm of the RPE.\(^9\)\(^,\)\(^10\)\(^,\)\(^12\)\(^-\)\(^14\)\(^,\)\(^18\)\(^-\)\(^22\)\(^,\)\(^28\) In contrast, our series of adenoma of the IPE (20 patients)\(^3\) and CPE (8 patients)\(^6\) revealed an equal sex distribution in each.

Our study has revealed new information about the clinical features and course of acquired tumors of the RPE. Although most are dark gray to black, they occasionally can be nonpigmented. In patient 6, the tumor was not the typical black and was believed clinically to be a vascular tumor or amelanotic choroidal melanoma, before FNAB and subsequent histopathologic examination of the enucleated globe confirmed the diagnosis of adenocarcinoma of the RPE.\(^24\) Both tumors in our series that were classified as adenocarcinoma were also nonpigmented, an observation for which we have no explanation.

There were 2 other interesting new observations in our series of RPE tumors. First was the observation of a prominent retinal artery and vein that supplied and drained the tumor. Such vessels were clearly visualized in 8 of our 13 patients. In fact, they were observed in every

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**Table 1. Clinical Data on 13 Patients With Acquired Neoplasms of the Retinal Pigment Epithelium**

<table>
<thead>
<tr>
<th>Patient No./ Sex/Age, y/ Race/Year of Diagnosis</th>
<th>VA Location</th>
<th>Size, mm</th>
<th>Associated Findings</th>
<th>Management</th>
<th>TNAB Cytologic Diagnosis</th>
<th>Pathologic Diagnosis</th>
<th>Comments and Follow-up, y</th>
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<tbody>
<tr>
<td>1/F/76/W</td>
<td>1980 6/6</td>
<td>JP</td>
<td>2 × 2 × 1</td>
<td>Observation</td>
<td>Adenoma</td>
<td>Stable</td>
<td>Postmortem examination</td>
</tr>
<tr>
<td>2/F/47/W</td>
<td>1988 6/6</td>
<td>P</td>
<td>9 × 7 × 4</td>
<td>Observation</td>
<td>Adenoma</td>
<td>Stable</td>
<td>10</td>
</tr>
<tr>
<td>3/F/58/W</td>
<td>1990 6/6</td>
<td>P</td>
<td>5 × 5 × 3</td>
<td>Observation</td>
<td>Adenoma</td>
<td>Stable</td>
<td>8</td>
</tr>
<tr>
<td>4/F/46/A</td>
<td>1991 6/21</td>
<td>MP</td>
<td>8 × 8 × 6</td>
<td>Enucleation</td>
<td>Adenoma</td>
<td>Stable</td>
<td>1</td>
</tr>
<tr>
<td>5/F/34/A</td>
<td>1992 6/12</td>
<td>MP</td>
<td>9 × 9 × 5</td>
<td>Plaque</td>
<td>Adenoma</td>
<td>Stable</td>
<td>NLP, 6</td>
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<tr>
<td>6/F/66/W</td>
<td>1992 6/50</td>
<td>JP</td>
<td>6 × 5 × 3</td>
<td>Histoplasmosis, RD</td>
<td>Adenocarcinoma Adenocarcinoma</td>
<td>Stable, 6</td>
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<tr>
<td>7/M/28/A</td>
<td>1993 HM MP</td>
<td>14 × 14 × 8</td>
<td>RFV, RD</td>
<td>Enucleation</td>
<td>Adenocarcinoma Adenocarcinoma</td>
<td>Stable, 6</td>
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<tr>
<td>8/F/54/W</td>
<td>1993 CF P</td>
<td>7 × 7 × 5</td>
<td>Cataract</td>
<td>Resection</td>
<td>Adenoma</td>
<td>Stable</td>
<td>6</td>
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<tr>
<td>9/M/54/W</td>
<td>1994 6/6</td>
<td>MP</td>
<td>4 × 4 × 4</td>
<td>RFV</td>
<td>Laser elsewhere</td>
<td>Stable</td>
<td>1</td>
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<tr>
<td>10/F/40/W</td>
<td>1994 6/120</td>
<td>P</td>
<td>6 × 5 × 3</td>
<td>RFV, macular hole</td>
<td>Adenoma</td>
<td>Stable</td>
<td>4</td>
</tr>
<tr>
<td>11/F/79/W</td>
<td>1995 NLP P</td>
<td>17 × 17 × 17 Staphyloma</td>
<td>Enucleation</td>
<td>Adenocarcinoma Stable, 3</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>12/M/66/W</td>
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<td>P</td>
<td>8 × 8 × 3</td>
<td>RFV, RD, ARM, SWR</td>
<td>FNAB, observation Adenoma</td>
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<tr>
<td>13/F/42/W</td>
<td>1995 6/120</td>
<td>M</td>
<td>6 × 6 × 4</td>
<td>RD, macular scar</td>
<td>Observation</td>
<td>Growth, 3</td>
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</table>

*VA indicates visual acuity; FNAB, fine needle aspiration biopsy; W, white; A, African American; JP, juxtapapillary; P, peripheral; MP, midperipheral; M, macula; RFV, retinal feeder vessel; SWR, surface wrinkling retinopathy; RD, retinal detachment; ARM, age-related macular degeneration; VH, vitreous hemorrhage; HM, hand motions; NLP, no light perception; CF, counting fingers; and ellipses, not done or information not available.
patient in whom the tumor was located in the peripheral fundus and in whom the ocular media were clear. It appears that tumors of the RPE have a tendency to invade the sensory retina. Curiously, the 3 tumors that produced the most pronounced dilated, tortuous retinal vessels all occurred in our African American patients.

The second apparent new observation in our patients was the occurrence of intraretinal exudation and exudative retinal detachment. This exudative process is similar to that seen with retinal capillary hemangioma and Coats disease. The combination of the tumor-associated retinal blood vessels and the exudative retinal detachment are remarkably similar to those seen with the retinal capillary hemangioma of von Hippel, except that the tumor is black, rather than red.

In patients 2 and 4, the RPE tumor induced vitreous hemorrhage rather than exudative retinopathy. We are uncertain why this pattern develops in some patients, but it is also similar to that which occurs in some patients with retinal capillary hemangioma, which also can assume a vitreoretinal or an exudative form. Other changes seen in our patients included surface wrinkling retinopathy and macular hole (Table 1).

Most reported tumors of the RPE were diagnosed clinically as melanoma, and the patient was treated with enucleation. Since tumors of the RPE have no potential for metastasis, it would be desirable to have clinical criteria for differentiating them from choroidal melanomas. Like choroidal melanoma, a neoplasm of the RPE is a pigmented fundus lesion of adulthood that grows slowly and can produce retinal detachment.

![Figure 1](https://example.com/figure1.png)

**Figure 1.** Clinical characteristics of tumors of the retinal pigment epithelium (RPE). A, Typical adenoma of the RPE located at the equator, showing abruptly elevated black mass. The camera is focused on the tumor surface, and the retinal feeder vessels are out of focus and cannot be visualized (patient 2). B, Another typical adenoma of the RPE located at the equator. In this instance, the camera is focused deeper on the adjacent retina, showing a minimally dilated feeder artery and drainer vein (patient 3). C, Pigmented adenoma of RPE partially over optic disc, simulating a melanocytoma (patient 4). D, Minimally pigmented adenocarcinoma of the RPE adjacent to optic disc with secondary intraretinal exudation. The diagnosis was confirmed after enucleation (patient 6). E, Fundus diagram of an adenoma of the RPE showing pigmented mass with dilated, tortuous retinal feeder vessels and extensive exudative retinal detachment in a 34-year-old African American woman (patient 8). F, Fundus photograph of lesion shown in part E depicting subretinal exudation and retinal feeder vessels coming from the disc (above) and leading to a deeply pigmented mass. The edge of the black tumor is seen down and to the right. G, Fundus photograph of entire tumor shown in part E showing abruptly elevated, dome-shaped mass with surrounding intraretinal and subretinal exudation. H, Fundus photograph of another adenoma of the RPE showing pigmented mass with dilated, tortuous retinal feeder vessels and extensive exudative retinal detachment in a 28-year-old African American man (patient 7). The optic disc is to the left, and the edge of the black mass is to the right.

![Figure 2](https://example.com/figure2.png)

**Figure 2.** Fluorescein angiographic features of adenoma of the retinal pigment epithelium. A, Arterial-phase angiogram of lesion shown in Figure 1, parts E to G, demonstrating dilated tortuous, feeding artery and hypofluorescence of the tumor. B, Venous-phase angiogram, showing continued relative hypofluorescence of the tumor with early leakage from the overlying retinal vessels.

![Figure 3](https://example.com/figure3.png)

**Figure 3.** Ultrasonographic features of adenoma of the retinal pigment epithelium. A, A-scan of lesion shown in Figure 1, parts E to G, demonstrating high internal reflectivity of the tumor. B, B-scan of lesion demonstrating abruptly elevated dome-shaped mass with acoustic solidity and secondary retinal detachment.
The RPE tumor is very likely to produce yellow intraocular or subretinal exudation, often leading to an exudative retinal detachment, similar to a retinal capillary hemangioma. Although a choroidal melanoma frequently produces a retinal detachment, the subretinal fluid usually is serous in nature, and appreciable yellow retinal or subretinal exudation is extremely rare in a comparably sized melanoma.

Another distinguishing feature of RPE adenomas is the development of a retinal blood supply. An RPE tumor is quite likely to exhibit a feeding retinal artery and a draining retinal vein. Eight of the RPE tumors had such vessels. Although a melanoma can rarely invade the retina and cause dilated tortuous retinal vessels, it is usually only the vein, and not the artery, that becomes dilated. Other conditions to be considered in the differential diagnosis include congenital hypertrophy of the RPE, combined hamartoma of the RPE, and congenital hyperplasia of the RPE. The characteristic features of these conditions are reported elsewhere, and they should be easily differentiated from the larger lesions reported herein.

Ancillary studies also can help in the differentiation of a neoplasm of the RPE from melanoma. With results of fluorescein angiography, the RPE tumor tends to be relatively hypofluorescent in the filling phases (Figure 2). Choroidal melanoma generally shows earlier hyperfluorescence and intense late staining. Also, an RPE tumor does not have the extensive internal vascularity and the double circulation that typifies many melanomas. Ultrasonography of a tumor of the RPE generally reveal high internal reflectivity with A-scan and an acoustically solid, abruptly elevated, dome-shaped mass with B-scan (Figure 3). In contrast, ultrasonography of a melanoma is more likely to reveal low internal reflectivity with A-scan and a sloping margin and acoustic hollowness with B-scan.

Another diagnostic procedure that can assist in the diagnosis is FNAB. Fine needle aspiration biopsy should generally be reserved for cases where there is diagnostic uncertainty and where a microscopic diagnosis is necessary to make a therapeutic decision. Diagnostic FNAB was performed in 4 of our 13 patients, and results showed cytologic findings compatible with RPE tumor in each instance. Results of cytologic examination of a tumor of the RPE demonstrate deeply pigmented, plump, round cells with large melanosomes, compared with the spindle

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Location</th>
<th>Procedure</th>
<th>Pathologic Diagnosis</th>
<th>Pattern</th>
<th>Pigment</th>
<th>Local Invasion</th>
<th>Nuclear Atypia‡</th>
<th>Basement Membrane‡</th>
<th>Prominent Tumor Vessels</th>
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<td>1</td>
<td>JP</td>
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<td>Adenoma</td>
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<td>3</td>
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<td>2</td>
<td>P</td>
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<td>Adenoma</td>
<td>Combined</td>
<td>3</td>
<td>Retina, vitreous</td>
<td>1</td>
<td>3</td>
<td>Yes</td>
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<td>4</td>
<td>MP</td>
<td>Resection</td>
<td>Adenoma</td>
<td>Combined</td>
<td>3</td>
<td>Retina</td>
<td>1</td>
<td>1</td>
<td>Yes</td>
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<td>JP</td>
<td>Resection</td>
<td>Adenocarcinoma</td>
<td>Solid</td>
<td>4</td>
<td>Retina</td>
<td>4</td>
<td>3</td>
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<td>Resection</td>
<td>Adenoma</td>
<td>Vacuolated</td>
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<td>Retina, uvea</td>
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<td>Resection</td>
<td>Adenocarcinoma</td>
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<td>0</td>
<td>Retina, uvea, orbit</td>
<td>3</td>
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<td>11</td>
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<td>0</td>
<td>Retina</td>
<td>3</td>
<td>3</td>
<td>No</td>
</tr>
</tbody>
</table>

*JP indicates juxtapapillary; P, peripheral; MP, midperipheral; and question mark, unknown.
†Graded on a scale of 0 to 4, with 0 indicating minimal involvement; 4, marked involvement.
cells and smaller melanosomes that characterize most aspirates of melanomas.

We obtained microscopic verification of the diagnosis of neoplasm of the RPE in 10 of our 13 patients. In the other 3, the typical clinical features, described herein, strongly supported the diagnosis. Interestingly, the histopathologic pattern is generally different in tumors of the posterior aspect of the RPE and CPE. Tumors of the posterior RPE tend to form more glandular or tubular elements, whereas those of the peripheral RPE and CPE tend to form vacuoles.

Tumors of the RPE are generally considered to be benign lesions that lack a tendency to metastasize. Nevertheless, we have shown that some tumors of the RPE can grow and cause profound visual loss. This aggressive nature of many tumors of the RPE raises questions about the best management. If a suspected tumor of the RPE is small and asymptomatic, it may be justifiable to observe the patient periodically and withhold treatment until visually threatening complications begin to develop. If the diagnosis is uncertain, FNAB may be a valuable method for confirming the diagnosis and excluding the possibility of melanoma. However, FNAB should be performed by experienced clinicians, and the aspiration should be studied by an experienced cytopathologist. For progressive or symptomatic lesions that are circumscribed and located in the peripheral fundus, local resection of the tumor is preferable. For more posteriorly located tumors, the role of laser photocoagulation and cryotherapy is not established, but such measures could be attempted in patients with early growth or visually threatening complications. Our small series suggests that tumors of the RPE are not particularly sensitive to irradiation, but plaque brachytherapy or charged particle irradiation can be attempted when there are no other legitimate options.

Our study has provided information about the clinical features and clinical course of acquired tumors of the RPE. Although these tumors can be stationary for a long time, many of them demonstrate progression, ocular complications, and visual loss. In contrast to choroidal melanomas, tumors of the RPE are likely to be dark black, abruptly elevated, and dome shaped; to assume a blood supply from the sensory retina; and to cause an exudative retinal detachment. We believe that the diagnostic guidelines set forth herein will help the clinician to diagnose tumors of the RPE and ultimately to provide better care of patients affected by this unusual neoplasm.

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