Advances in chemotherapy involving cisplatin-based regimens markedly improved the prognosis in the mid-1970s. In fact, germ cell tumors have become one of the most curable solid neoplasms, with survival rates of approximately 80% for advanced disease and nearly 100% for early disease.2

The visual prognosis in metastatic choriocarcinoma to the choroid has been poor. Most patients described prior to the 1970s died without any improvement in their vision. In the 2 cured patients, one’s visual acuity was not reported,3 whereas the other’s was 20/1000 due to scarring from the regressed tumor.4 Despite the relative afferent pupillary defect and the large tumor mass (12.4 mm), our case had an excellent visual outcome of 20/40, highlighting the importance of close and continued ophthalmic care. During chemotherapy, serial ultrasonography examinations were essential in showing the treatment response by documenting the decrease in tumor size. Ophthalmologists must be patient with the chemotherapy before considering surgical therapy, although surgical options should still be considered after treatment if appropriate. Close follow-up in conjunction with an oncologist is required.

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Perivascular Epithelioid Cell Tumor of the Orbit

Perivascular epithelioid cell tumors (PEComas) are rare mesenchymal neoplasms comprising angiomylipoma, lymphangiomyomatosis, and clear cell “sugar” tumor of the lung.1 In 1992, the term PEC was introduced to characterize distinct HMB45-positive cells that seemed to originate from blood vessel walls.2 Zamboni et al3 coined the term PEComa to describe a pancreatic clear cell sugar tumor lesion indistinguishable from lung clear cell sugar tumor. Recently, PEComas have been recognized at various sites, most often in middle-aged female patients.3 PEComas are characterized by typical chromosomal imbalances, suggesting PECs as distinct tumor cells.4

A single case of orbital PEComa has been reported in a 9-year old child.5 Here we report a PEComa in the orbit of a 54-year-old patient.

Report of a Case. A 54-year-old male patient had a slowly progressing, painless swelling of the right temporal lower eyelid (Figure, A). Ophthalmological examination results of the healthy patient were otherwise normal. Orbital examination showed a soft fluctuant mass in the anterior inferotemporal orbit without overlying cutaneous changes. Orbital ultrasonography and computed tomography revealed a highly reflective, demarcated, hypodense, contrast-enhancing, round lesion measuring 1.5 × 1.0 × 1.0 cm. The tumor with large feeder vessels was completely excised through an anterior orbitotomy with subciliary incision. No recurrence was detected during 17 months of follow-up.

Figure 2. Fundus photographs. In the periphery, fundus photographs show retinal pigment epithelial changes with subretinal fibrosis in the area of previously active metastatic lesions.

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The tumor was well delineated with a yellow cut surface (Figure, B). Light microscopy revealed a thin fibrous capsule and lymphocytic infiltrates (Figure, C). Solid and trabecular tumor cell aggregates showed intimate association with a ramified vascular network (Figure, D). The tumor cells were large with a clear to eosinophilic granular cytoplasm, round to oval nuclei with prominent nucleoli, and diffuse cell borders. Few cells contained melanin.

The melanocytic markers HMB45 and MART1 (melan A) (Figure, E) were strongly expressed, as were CD10 and CD68. Staining results for the pancytokeratin marker AE1/3, desmin, S-100 protein, CD31, actin, CD117, microphthalmia transcription factor, and chromogranin were negative. Vimentin stained single large cells likely not belonging to the tumor. The proliferation index (ki67) was very low (<1%).

**Comment.** PEComas are composed of epithelioid, sometimes spindled cells with clear to eosinophilic granular cytoplasm in association with blood vessel walls. Characteristic features are HMB45 and MART1 expression, variable expression of muscle markers, and no cytokeratin or S-100 protein expression. In a recent comprehensive review, expression patterns were as follows: HMB45, 100% of PEComas; MART1, 41%; smooth muscle actin, 59%; S-100 protein, 11%; desmin, 31%; cytokeratin, 0%; and CD117, 33%. In our case, we detected melanocytic markers but no epithelial or smooth muscle markers.

The differential diagnosis of clear cell tumors is vast and comprises clear cell tumors of the lung, kidney, or female genital tract, which can be ruled out in the absence of epithelial markers.

As PEComas may produce melanin, malignant melanoma and pigmented paraganglioma are important differential diagnoses. Orbital melanoma is rare and includes primary orbital melanoma, secondary orbital melanoma by extension from adjacent structures, and metastatic melanoma. PEComas expressing S-100 protein usually also stain for smooth muscle antigen. In our case, the lack of nuclear polymorphism or S-100 protein staining and low mitotic activity make the diagnosis of melanoma unlikely. The epithelioid cell structure resembles paraganglioma, but the lack of neurosecr-
tory granules or staining for chromogranin and S-100 protein argue against it.

PEComas generally show a benign course, although rare cases of malignant PEComas have been published. Necrosis, a high mitotic rate, a size larger than 7 cm, an infiltrative growth pattern, and a remarkable hypercellularity may indicate malignant PEComa. These characteristics were absent in our specimen. Because of its rarity, the clinical features, biological behavior, and best treatment of orbital PEComa are currently unclear. As most PEComas are benign, primary excision and regular follow-up seem reasonable.

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Archives Web Quiz Winner

Congratulations to the winner of our February quiz, Anmar M. Abdul-Rahman, FRANZCO, MOphth, Department of Ophthalmology, Middlemore Hospital, Auckland, New Zealand. The correct answer to our February challenge was paraneoplastic retinopathy. For a complete discussion of this case, see the Letters: Research Letters section in the March Archives (Eksandh L, Adamus G, Mosgrove L, Andréasson S. Autoantibodies against bestrophin in a patient with vitelliform paraneoplastic retinopathy and a metastatic choroidal malignant melanoma. Arch Ophthalmol. 2008;126(3):432-435).

Be sure to visit the Archives of Ophthalmology Web site (http://www.archophthalmol.com) and try your hand at our Clinical Challenge Interactive Quiz. We invite visitors to make a diagnosis based on selected information from a case report or other feature scheduled to be published in the following month’s print edition of the Archives. The first visitor to e-mail our Web editors with the correct answer will be recognized in the print journal and on our Web site and will also be able to choose one of the following books published by AMA Press: Clinical Eye Atlas, Clinical Retina, or Users’ Guides to the Medical Literature.

Author Contributions: Dr R. Guthoff had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

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Additional Contributions: Christopher D. M. Fletcher, FRCPath, confirmed the diagnosis.