Improved Systemic Chemotherapy for Metastatic Testicular Choriocarcinoma Can Result in Excellent Prognosis for Life and Vision

Although testicular germ cell carcinoma is rare, it is the most common malignant neoplasm in men between the ages of 15 and 35 years. We report a case of metastatic choriocarcinoma involving the choroid that was effectively treated with chemotherapy and had an excellent visual outcome.

Report of a Case. A 23-year-old healthy man visited the emergency department with decreased vision in his left eye. His best-corrected visual acuities were 20/20 OD and light perception OS. There was a left afferent pupillary defect. Slitlamp and fundus examination results of the right eye were unremarkable. Dilated funduscopic examination of the left eye was limited due to a vitreous hemorrhage. Standardized A-scan and contact B-scan ultrasonography of the left eye showed a large, irregularly shaped and structured, highly vascular mass lesion in the superotemporal quadrant (Figure 1). The mass measured approximately 19.0 × 15.8 mm in diameter with a maximal thickness of 12.4 mm.

On further detailed review of systems, the patient admitted to a 6-week history of testicular pain. An initial workup revealed a testicular mass, and chest radiography revealed 2 mass lesions in the lungs. The patient was immediately referred to an oncologist for further metastatic workup. The patient was diagnosed with metastatic testicular choriocarcinoma requiring radical orchiectomy followed by a 5-cycle chemotherapy regimen of bleomycin sulfate, etoposide, and cisplatin.

The patient responded well to the chemotherapy systemically, and serial ultrasonography examinations showed a dramatic decrease in the thickness of the choroidal mass from 12.4 mm to 3.0 mm. Three months after chemotherapy, the patient was clinically in remission. The choroidal mass continued to decrease in size until it became undetectable by ultrasonography (Figure 1) and the left afferent pupillary defect resolved. However, the visual acuity was 6/200 OS from a nonclearing vitreous hemorrhage. Five months into remission without resolution of the hemorrhage, the patient underwent a 25-gauge pars plana vitrectomy of the left eye. He later developed an epiretinal membrane and cataract that required a vitrectomy with membrane peeling and cataract extraction. Three years after his initial diagnosis, the patient remains in remission with a visual acuity of 20/40 OS. Subretinal fibrosis has replaced the previously active metastatic lesions (Figure 2).

Comment. Choriocarcinoma is an extremely rare cause of choroidal metastasis. A recent meta-analysis reported 15 cases of testicular choriocarcinoma to the choroid. Seventy-three percent of these patients (11 of 15 patients) had a poor prognosis, with survival of less than 2 months in the majority of these cases. Most of these reports with poor survival were prior to the 1970s. Ad-
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.

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, whereas the other’s was 20/1000 due to scarring from the regressed tumor. 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, lymphangioleiomyomatosis, and clear cell “sugar” tumor of the lung. In 1992, the term PEC was introduced to characterize distinct HMB45-positive cells that seemed to originate from blood vessel walls. Zamboni et al 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. PEComas are characterized by typical chromosomal imbalances, suggesting PECs as distinct tumor cells.

A single case of orbital PEComa has been reported in a 9-year old child. 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, hyperdense, 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.