Case Report

Choroidal metastasis from leiomyosarcoma in two cases

Eric Feinstein, Swathi Kaliki[1], Carol L. Shields[2], Hormoz Ehya[3], Jerry A. Shields[2]

Department of Ophthalmology and Visual Sciences, University of Illinois-Chicago, Chicago, IL USA, [1]Ocular Oncology Service, L V Prasad Eye Institute, Hyderabad, India, [2]Ocular Oncology Service, Wills Eye Hospital, Thomas Jefferson University Philadelphia, [3]Department of Pathology, Fox Chase Center, Philadelphia, PA, USA,

Leiomyosarcoma is a malignant tumor of mesenchymal cells and is the most common soft-tissue sarcoma. Leiomyosarcoma is a notably rare tumor in the ophthalmic region and can be of primary, secondary or metastatic origin. To the best of our knowledge, there has only been one published case of leiomyosarcoma metastasis to the choroid. In this case study, we report two cases of primary leiomyosarcoma with metastasis to the choroid of the eye. Both cases displayed systemic metastasis and showed response to high dose plaque radiotherapy. Despite its prevalence as the leading form of sarcoma, leiomyosarcoma rarely metastasizes to the ocular region.

Keywords: Choroid, eye, fine needle aspiration biopsy, leiomyosarcoma, metastasis, sarcoma, tumor, uvea

Introduction

Leiomyosarcoma is a malignant tumor of mesenchymal cells with smooth muscle differentiation and represents the most common soft-tissue sarcoma. In a population-based cancer incidence review of 26,758 cases of soft-tissue sarcoma, the leading types included leiomyosarcoma (24%), malignant fibrous histiocytoma (17%), liposarcoma (11.5%) and dermatofibrosarcoma (10.5%).[1] Leiomyosarcoma can arise from various sites including soft-tissue (48%), skin (14%), gastrointestinal tract (7%), uterus (7%) and retroperitoneum (7%).[1]

Metastasis from leiomyosarcoma depends on the site of origin. Cutaneous leiomyosarcoma rarely metastasizes, whereas retroperitoneal and large blood vessel leiomyosarcoma display metastatic rate of 40-50%.[2] Distant metastasis occurs generally by hematogenous route to liver (53%) and lung (47%).[2] Metastasis from leiomyosarcoma to the ocular region is extremely rare. In a comprehensive survey of 950 metastatic tumors to the eye, there were no cases of metastasis from leiomyosarcoma.[3] Furthermore, PubMed search for keywords "leiomyosarcoma," "choroid," "metastasis," "uvea" and "eye" produced only one documented case of intraocular metastasis from leiomyosarcoma. Herein, we report two cases of leiomyosarcoma with intraocular metastasis.

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Case Reports

Case 1

A 66-year-old Caucasian male had a 4-month history of blurred vision in the left eye oculus sinister (OS). He had a retroperitoneal leiomyosarcoma surgically excised 4 years earlier with known liver and lung metastasis that were treated with systemic chemotherapy. Visual acuity (VA) was 20/25 in the right eye (oculus dexter) and 20/50 OS. Ophthalmoscopy disclosed a solitary, juxtapapillary yellow-white choroidal tumor measuring 5 × 3 mm in diameter [Figure 1]. Ultrasonography OS revealed a hollow, dome-shaped, 2.9 mm thick choroidal mass. Optical coherence tomography confirmed subfoveal fluid. Transvitreal fine needle aspiration biopsy (FNAB) yielded only a few atypical spindle cells, which were inadequate for confirmatory immunocytochemical studies. Treatment with photodynamic therapy (PDT) did not control the tumor and thickness increased to 3.2 mm, so iodine-125 (I^{125}) plaque radiotherapy (8000 cGy apex dose) was provided with subsequent tumor control. Radiation maculopathy lead to reduced VA of 20/100 OS. Patient succumbed from systemic metastasis 9 months following ocular treatment.

Case 2

A 62-year-old Caucasian male noted peripheral visual field defect OS for 4 weeks. He had a history of pulmonary leiomyosarcoma

Correspondence:
Dr. Carol L. Shields, Ocular Oncology Service, Suite 1440, Wills Eye Institute, 840 Walnut Street, Philadelphia, PA 19107, USA. E-mail: carol.shields@shieldsoncology.com

surgically excised 4 years previously and with known groin and buttocks metastasis that were surgically excised and shoulder and throat metastasis that were treated with external beam radiotherapy. VA was 20/25 both eyes (OU). Ophthalmoscopy revealed a solitary, superonasal, ciliochoroidal mass, 13.7 mm in basal diameter [Figure 2a]. Ultrasonography disclosed a mushroom-shaped choroidal mass measuring 9.7 mm in thickness and with subretinal fluid [Figure 2b and c]. FNAB of the mass showed clusters of atypical spindle cells with elongated, hyperchromatic nuclei and irregular nuclear membrane [Figure 2d]. Occasional small nucleoli and mitotic figures were observed [Figure 2e]. Although the material was inadequate for immunocytochemical studies, the cytomorphologic features were consistent with metastatic leiomyosarcoma. Treatment with I^{125} plaque radiotherapy (8000 cGy apex dose) was performed. Tumor reduction to 6 mm thickness was achieved by 15 months

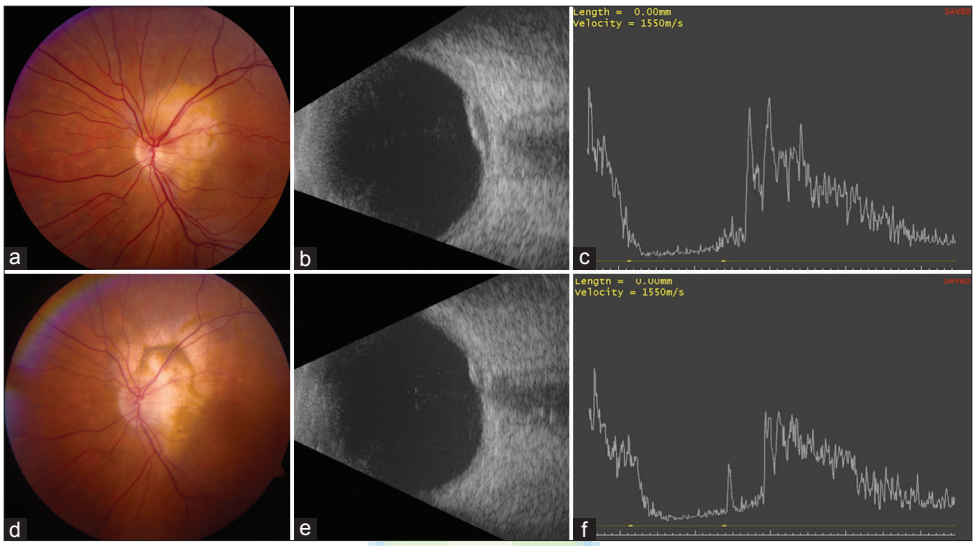

Figure 1: A 66-year-old Caucasian male with juxtapapillary choroidal metastasis (a-c) before treatment and regressed tumor (d-f) after plaque radiotherapy

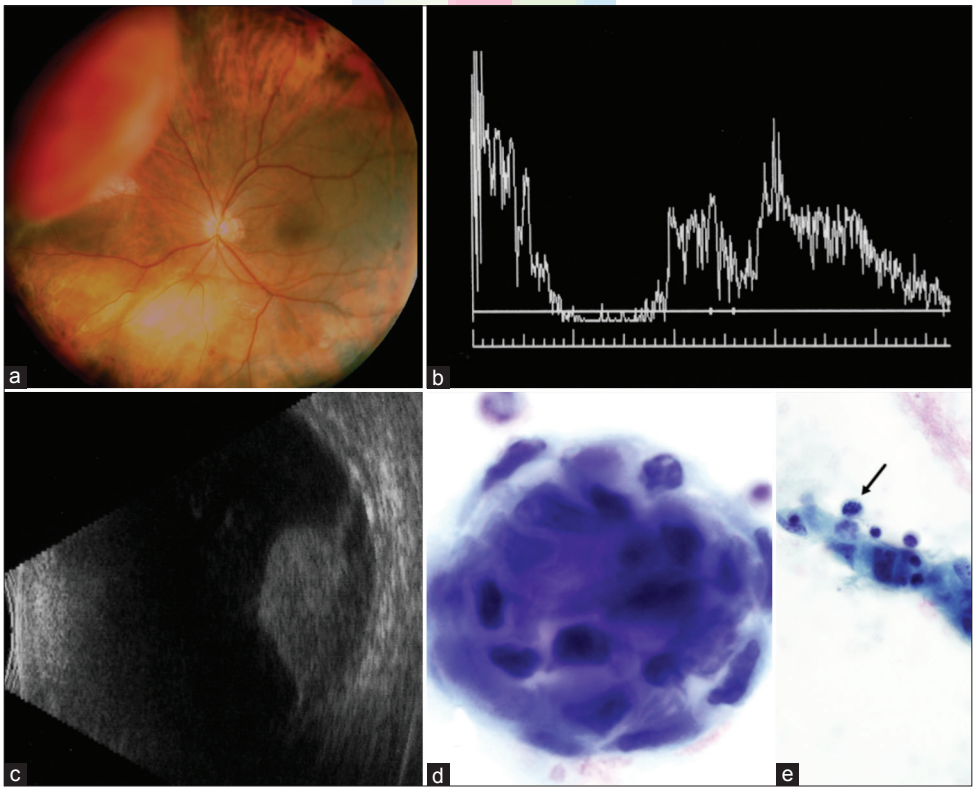

Figure 2: A 62-year-old Caucasian male with superonasal equatorial choroidal metastasis (a), with high to medium internal reflectivity on A scan (b) and mushroom shaped configuration on B-scan ultrasonography (c). Fine needle aspiration biopsy yielded a cohesive cluster of atypical spindle cells with hyperchromatic, elongated nuclei (d) and a mitotic figure (black arrow) (e) (Papanicolaou stain). Following plaque radiotherapy, tumor regression was documented but persistent vitreous hemorrhage and retinal detachment lead to poor visual acuity

follow-up. Persistent retinal detachment and vitreous hemorrhage lead to reduced VA of counting fingers.

Discussion

Leiomyosarcoma is a notably rare tumor in the ophthalmic region and can be of primary, secondary or metastatic origin. Primary ophthalmic leiomyosarcoma, found rarely in the orbit, is believed to develop from vascular or sympathetic smooth muscle.[4] Secondary, leiomyosarcoma has been documented to arise following radiation therapy for retinoblastoma.[4] Metastatic leiomyosarcoma to the orbital region has been identified from primary tumors in the uterus, gastrointestinal tract and soft-tissues.[4] To the best of our knowledge, only one previous case report of choroidal metastasis from leiomyosarcoma was found on PubMed.[5] In that case, a 22-year-old woman underwent radiotherapy then resection of a large choroidal metastasis that proved on immunohistochemistry to represent leiomyosarcoma from a primary site in the lung.

Shields *et al*. conducted a retrospective study on 950 uveal metastasis in 520 eyes of 420 consecutive patients, 88% of which were located within the choroid.[3] Uveal metastasis originated from primary cancer of the breast (47%), lung (21%), gastrointestinal tract (4%), kidney (2%), skin (2%) and prostate (2%) while the primary site remained unknown in 17% of cases despite systemic evaluation.[3] The primary tumors were found to be carcinomas (82%), unknown origin (13%), melanoma (3%), carcinoid (1%) and sarcoma (<1%) (Ewing's sarcoma).[3] There was no case of metastatic leiomyosarcoma.

Choroidal metastasis appears as a dome or plateau shaped yellow mass, often with secondary retinal detachment and most have co-existing systemic metastasis.[3] Both cases in our series had coexistent systemic metastasis. Treatment for choroidal metastasis includes observation for regressed lesions, chemotherapy, hormone therapy, external beam radiotherapy, plaque radiotherapy or local resection for active lesions.[6] PDT is an emerging option. Case 1 was initially treated with PDT, but failed and plaque radiotherapy was performed. Plaque radiotherapy can be an effective therapy, but leiomyosarcoma is somewhat radioresistant, hence higher dose radiotherapy was employed.[6] In our cases, both tumors showed regression.

In summary, we report two cases of leiomyosarcoma metastatic to the choroid. Both displayed systemic metastasis and showed response to high dose radiotherapy. Despite its prevalence as the leading form of sarcoma, leiomyosarcoma rarely metastasizes to the ocular region.

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