Metastatic leiomyosarcoma to the choroid

Leiomyosarcoma is the most common form of soft-tissue sarcoma arising from smooth muscle cells.[1] Metastasis of leiomyosarcomas typically occurs hematogenously,[2,3] with metastasis to the ocular region being extremely rare.[4,5]

A 70-year-old male presented with a 3-month history of blurry vision OD. The patient denied flashes, floaters, shadows, photophobia, diplopia, pain with eye movements, or a history of ocular trauma. Past medical history was significant for high-grade bone leiomyosarcoma of the left tibia first diagnosed 10 years ago that was surgically excised without adjuvant treatment. After 9 years of remission, the patient was found to have leiomyosarcoma metastasic to a rib on the right side (confirmed with biopsy), vertebrae, and lungs immediately preceding presentation to the ophthalmology clinic. Medications included pazopanib, zopiclone, and trazodone. Ocular and family histories were unremarkable. The patient did not smoke or drink alcohol.

On examination, the patient's visual acuity was 20/30 OD and 20/25 OS, with normal intraocular pressures. Both pupils were equal and reactive to light with no relative afferent pupillary defect. No proptosis, eyelid edema, chemosis, or extraocular muscle abnormalities were noted. On slit-lamp examination, the anterior segment was normal bilaterally. A dilated fundus examination of the right eye revealed an elevated subretinal mass with overlying retinal detachment approximately 1 disc diameter from the macula (Fig. 1) with the left eye normal. The differential diagnoses included metastasis, melanoma, and lymphoma.

Optical coherence tomography confirmed a choroidal mass in the temporal aspect of the right eye with a serous retinal detachment but no macular involvement. Optical coherence tomography of the left eye was normal. A B-scan showed a temporal dome-shaped mass 4.6 mm in elevation of nonhomogeneous moderate to high reflectivity OD. A Humphrey 30-2 visual field showed a unilateral nasal hemianopia OD, while the visual field was unremarkable OS. Fluorescein angiography OD showed early-phase hypofluorescence and late-phase heterogeneous hyperfluorescence in the temporal mass, while the fluorescein angiography OS was normal.

Orbital magnetic resonance imaging with contrast material showed a broad-based ocular lesion in the posterolateral aspect of the right globe measuring 4×9 mm and arising from the choroid without scleral involvement demonstrating intermediate T_1 and low to intermediate T_2 signal with homogeneous postcontrast enhancement (Fig. 2). The left globe was unremarkable. The most likely diagnosis was metastasis of leiomyosarcoma to the choroid of the right eye. The patient was referred to the radio-oncology service and underwent intensity-modulated stereotactic radiation therapy (IMRT) with a total dose of 30 Gy in 10 daily fractions over 2 weeks with protection of the surrounding tissue. Following 2 rounds of IMRT, the patient's cancer prognosis was deemed poor, IMRT was stopped, and patient was treated palliatively and passed away.

Leiomyosarcoma is a very rare ocular malignancy.[2,4] Ocular leiomyosarcoma is found rarely in the orbit and has been reported to be of vascular or sympathetic smooth muscle origin.[2] In a survey of uveal metastases over 20 years, Shields et al.[5] conducted a retrospective study of 950 uveal metastases, and 88% metastasized to the choroid. These uveal metastases originated primarily from the primary breast (47%), lung (21%), and gastrointestinal tract, kidney, skin, and prostate ($\leq$4%) cancers with no cases of metastatic leiomyosarcoma.[2,5] Metastatic leiomyosarcoma to the orbital region has been reported in the context of primary tumours from the buttock soft tissues, respiratory system, gastrointestinal system, and gynecologic system.[2,6]

We report a case of leimyosarcoma metastatic to the choroid in a patient with confirmed primary leiomyosarcoma of the left tibia. A review of the literature to date reveals only 3 other cases of leiomyosarcoma metastatic to the choroid reported globally.[2,7] Similar to our report, in the case published by Nieto et al.[7] and the 2 cases published by Feinstein et al.,[2] choroidal metastasis of leiomyosarcoma presented as a creamy white mass commonly associated with subretinal fluid, which has been shown to be the case in 28%−73% of patients.[5,9] Most choroidal metastases were posterior to the equator (80%).[5,8,9] In the case published by Nieto et al.,[7] anatomopathology or brachytherapy was not performed because of refusal by the patient, and ultimately, the patient was treated with trabectedin chemotherapy that failed after nearly 1 month with subsequent enucleation performed, and follow-up studies showed no recurrence or metastases. In the 2 cases of choroidal metastasis from leiomyosarcoma reported by Feinstein et al.,[2] 1 was a choroidal metastasis secondary to retroperitoneal leiomyosarcoma, and the other was secondary to pulmonary leiomyosarcoma. Transvitreal fine-needle aspiration biopsy for the first case yielded an inadequate sample and for the second case yielded clusters of atypical spindle cells with elongated, hyperchromatic nuclei and irregular nuclear membranes. Both patients responded to treatment with plaque radiotherapy for subsequent tumour control.

Treatment for choroidal metastasis is based on patient factors, prognosis, and type of cancer while balancing the risks and benefits.[2] External beam radiation therapy delivered with three-dimensional conventional radiotherapy techniques, intensity-modulated fractionated stereotactic radiotherapy (as in the case of our patient), single-fraction radiosurgery, proton therapy, and brachytherapy are the most common treatments for choroidal metastasis.[2,8,10] Other therapies include observation for regressed lesions, chemotherapy, hormone therapy, intravitreal injection, photodynamic therapy, and local resection for active lesions

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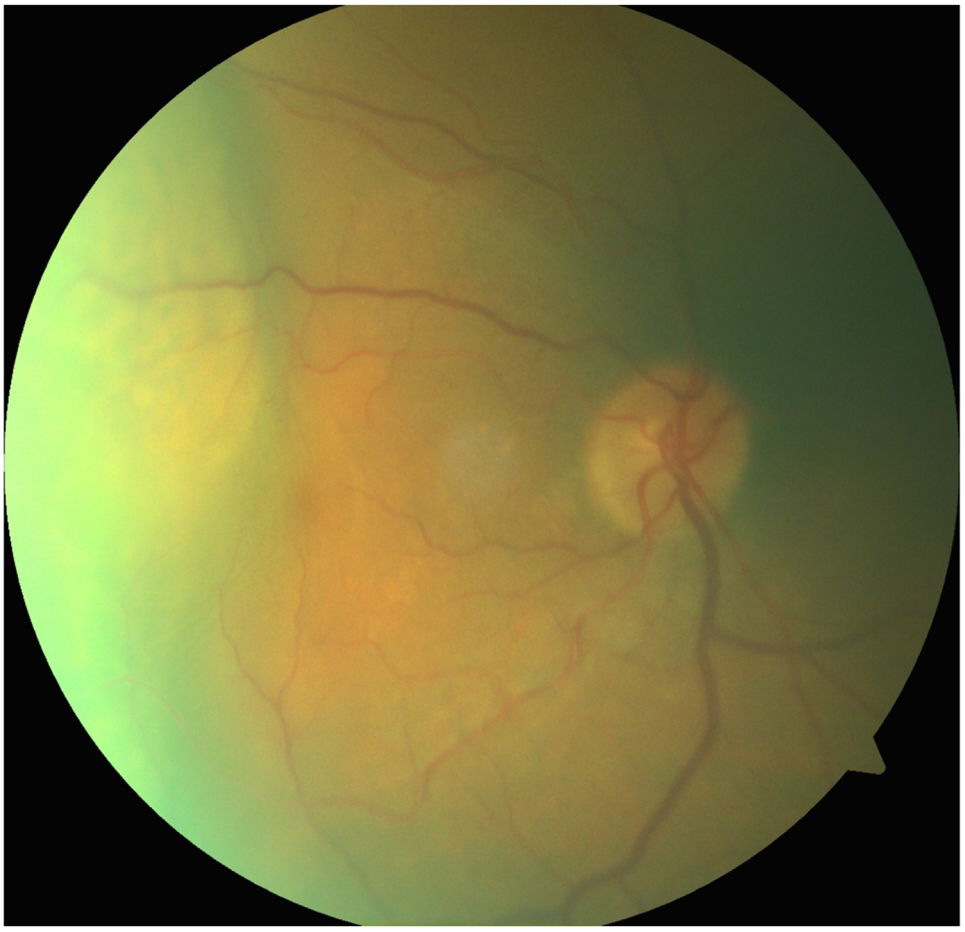

Fig. 1—Fundus photograph of the right eye at presentation showing a temporal raised subretinal mass from approximately 10 to 6 o'clock less than half a disc diameter from the macula.

in the orbit with mixed results.[2,8] Dose distribution, treatment planning, and cost vary widely among these techniques. The aims of radiotherapy are not only to control tumour growth but also to preserve vision by sparing radiation-sensitive areas (e.g., the macula and the optic disc).[8] During orbital radiation therapy, risks of radiation to other

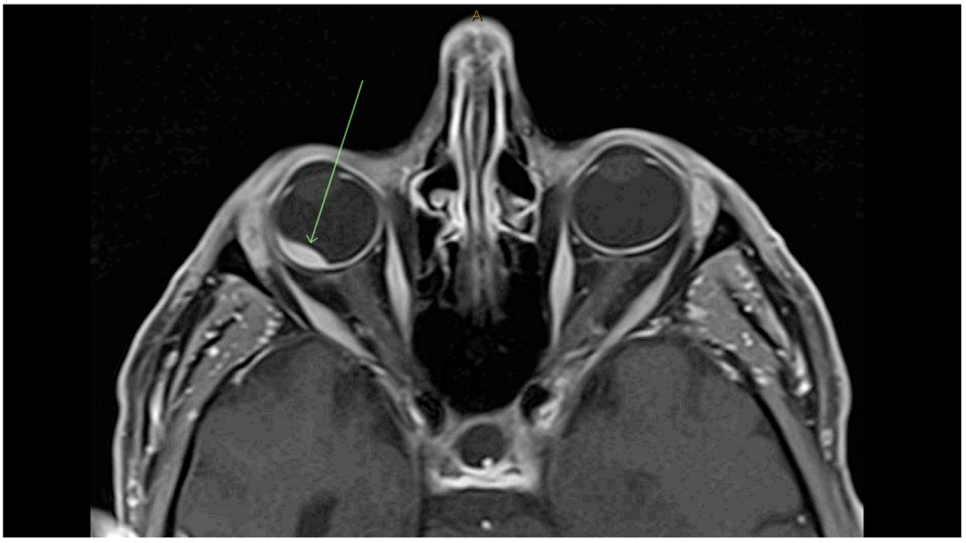

Fig. 2—Orbital magnetic resonance image with contrast material showing a broad-based ocular lesion in the posterolateral aspect of the right globe measuring 4 × 9 mm and arising from the choroid without scleral involvement demonstrating intermediate T_1 and low to intermediate T_2 signal with homogeneous postcontrast enhancement.

structures include the lens, cornea, retina, optic nerve, and optic chiasma.[8] External-beam radiation therapy and intensity-modulated radiation therapy for choroidal metastases appear to yield good outcomes, but there is limited evidence because no randomized studies have been published and only retrospective studies have been completed.[8,11]

In summary, we report a rare case of metastatic leiomyosarcoma to the choroid. Further research on treatment is needed.

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Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this correspondence.