Ultrasound Biomicroscopy in Management of Malignant Iris Melanoma

High-frequency ultrasound biomicroscopy (UBM) provides high-resolution cross-sectional images of anterior segment structures. Few published reports have correlated abnormal UBM images with histopathologic findings. A recent report by Marigo et al1 compared ultrasound images of iris and ciliary body melanomas with their histopathologic features. They found that UBM accurately defined the tumor shape and the extent of local invasion. There is histopathologic evidence that iris melanomas can spread through unguarded (Scheie procedure2 and iridencleisis3) and guarded (trabectectomy4) glaucoma procedures. We show that UBM can accurately determine the depth of tissue penetration by an iris melanoma into a trabeculectomy site by correlating the clinical images with the histopathologic features after enucleation.

Report of a Case. In January 1995, a 59-year-old white woman was evaluated for rising intraocular pressure in the left eye. The patient had been prescribed timolol maleate, pilocarpine, apraclonidine hydrochloride, and acetazolamide by the referring ophthalmologist. She had a history of a pigmented iris tumor in the left eye that had been stable for many years.

On examination, visual acuity was 20/20 OD and 20/30 OS. Applanation pressure was 15 mm Hg OD and 49 mm Hg OS. Results of slitlamp and gonioscopic examinations of the right eye were normal. The trabecular meshwork had minimal pigmentation. In the left eye, a slightly elevated pigmented lesion was visible in the peripheral iris between the 2- and 3-o'clock positions. Transillumination did not show involvement of the ciliary body. The angle was open, but clumps of pigment were layered in the inferior angle, and the entire trabecular meshwork was darkly pigmented. Results of a retinal examination in each eye were normal.

It was recognized that this iris lesion most likely was a malignant melanoma, but after extensive discussion, the patient chose to have a trabeculectomy to control intraocular pressure and preserve vision rather than undergo an immediate enucleation. The procedure was performed without complication. The postoperative course was uneventful, and intraocular pressure was reduced to 10 to 12 mm Hg without medications (Figure 1A).

The left eye continued to do well, with a visual acuity of 20/20 and an intraocular pressure of less than 20 mm Hg, until April 1996. At that time, the patient sought treatment for a 3-day history of eye pain; intraocular pressure was 32 mm Hg OS. Slitlamp examination showed patchy pigmentation over the entire iris surface and within the bleb cavity (Figure 1B). High-frequency UBM of the bleb and adjacent structures was performed with an Ultrasound Biomicroscope Model 840 (Zeiss Humphrey Systems, Dublin, Calif). This instrument has a frequency of 50 MHz and a lateral and axial resolution of 50 µm. The penetration depth is 5 mm. It scans at a rate of 8 Hz with a sampling resolution of 5 µm. Ultrasound biomicroscopy showed evidence of tumor proliferation within the sclerectomy and filtration bleb (Figure 2). The eye was enucleated. Results of a metastatic

Figure 1. A, A postoperative slitlamp photograph of the left eye shows a localized pigmented iris lesion between the 2- and 3-o’clock positions, a peripheral iridectomy at the 11:30-o’clock position, and a filtration bleb in the superonasal quadrant. B, Eleven months later, there is diffuse involvement of the iris and pigment in the filtration bleb.
workup, including a chest radiograph, liver function tests, and a computed axial tomographic scan of the abdomen and pelvis, were normal. Seven years later, the patient was in good health and without evidence of metastatic disease.

Pathologic Description. Gross examination of the enucleated specimen showed a $23 \times 23 \times 25$-mm left eye with 8.0 mm of optic nerve attached. A bleb in the superonasal quadrant measured $10 \times 6$ mm and an iris defect was present at the 11:30-o’clock position. A pigmented tumor arose from the peripheral iris and extended into the angle. Histopathologic analysis showed a diffuse melanoma of the iris that was predominately composed of spindle B cells but with a moderate number of epithelioid cells. Tumor infiltrated the trabecular meshwork, filtering bleb, and sclera adjacent to the operative site (Figure 3). There was seeding into the anterior chamber, on the trabecular meshwork, onto the anterior and posterior lens capsule and suspensory ligaments, and over the inferior pars plana. No extrascleral or extraconjunctival extension was seen. The optic nerve showed glaucomatous cupping with cavernous degeneration posterior to the lamina cribosa.

Comment. Iris melanomas are uncommon. In general, they grow slowly, and patients have an excellent prognosis because of their low metastatic potential (<5%). It is difficult to decide what course of action is appropriate when a pigmented iris lesion grows because benign and malignant lesions can enlarge and spread into adjacent structures. Both can cause elevated intraocular pressure. Because iris melanomas are more likely to metastasize if there is involvement of the iris root or angle with elevated intraocular pressure or when there is extraocular spread, it would be helpful to have a noninvasive clinical tool to help guide management decisions. In our case, it was remarkable how closely the findings from UBM mirrored the findings demonstrated by low-power microscopy of the fixed tissue. This study and others show the utility of high-frequency UBM in providing useful information about tumor morphology, growth pattern, and depth of penetration.

John R. Nordlund, MD
Neenah, Wis
Dennis M. Robertson, MD
David C. Herman, MD
Rochester, Minn

The authors have no relevant financial interest in this article.
Corresponding author and reprints: Dennis M. Robertson, MD, Department of Ophthalmology, Mayo Clinic, 200 First St SW, Rochester, MN 55905 (e-mail: robertson.dennis@mayo.edu).


Klippel-Trenaunay Syndrome and Rhabdomyosarcoma in a 3-Year-Old

Klippel-Trenaunay syndrome (KTS) is a congenital vascular anomaly with soft tissue and skeletal hypertrophy. It has been associated with capillary, venous, lymphatic, and soft tissue malformations, but not with malignancies. Orbital rhabdomyosarcoma typically demonstrates a rapid-onset orbital process that can be confused with trauma or benign tumors. This case report underscores that rhabdomyosarcoma can mimic a benign, lymphatic malformation, particularly in the setting of expected vascular lesions. A comprehensive MEDLINE search failed to identify a previous case of concurrent KTS and orbital rhabdomyosarcoma. Current chemotherapeutic regimens are reviewed.

Report of a Case. A 3-year-old boy had an inferonasal conjunctival mass of 10 days’ duration and a recent upper respiratory tract infection (URTI), including nasal congestion, rhinorrhea, and epiphora. Examination revealed a right, vascular conjunctival mass (Figure 1) and a grade 1 port-wine stain (diffuse capillary and venular malformation) of the left upper extremity (Figure 2A), with soft tissue hypertrophy of the left thenar eminence (Figure 2B). Computed tomography demonstrated an orbital mass with faint areas of internal septation (Figure 3).

The patient was referred to the multidisciplinary Hemangioma and Vascular Malformation Clinic at the

Figure 1. Clinical features of the right inferonasal conjunctival mass.

Figure 2. Clinical features of Klippel-Trenaunay syndrome. A, Reticulated pink mottling of the left chest and arm, consistent with a diffuse capillary malformation. B, Hypertrophied, hypervascular left thenar eminence in this right-handed boy.