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Case report

Uveal melanoma incidentally diagnosed with neuroimaging, a case series of 3 patients ☆

Mai Tsukikawa, MD[a,*], *Babatunde Akinpelu, MD*[b], *Pattana Wangaryattawanich, MD*[b], *Kathryn Scherpelz, MDPhD*[c], *Andrew W. Stacey, MDMS*[a]

[a] *Department of Ophthalmology, University of Washington, 325 Ninth Ave, Box 356421, Seattle, WA 98104, USA*
[b] *Department of Radiology, University of Washington, Seattle, WA, USA*
[c] *Department of Laboratory Medicine and Pathology, University of Washington, Seattle, WA, USA*

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ABSTRACT

Uveal melanoma is the most common primary intraocular malignancy and can occur in the choroid, the ciliary body, or the iris. It is most often diagnosed based on clinical examination by an ophthalmologist. Nearly all patients present with visual symptoms. Characteristic findings on clinical examination include pigmented or pale choroidal masses with serous retinal detachments and acoustic hollowness seen with ocular ultrasonography. CT and MRI of the orbits are not traditionally utilized for the diagnosis of uveal melanoma. We present 3 cases in which uveal melanoma was an incidental finding on neuroimaging for unrelated conditions in asymptomatic patients. Radiologists should maintain a high suspicion for uveal melanoma when an intraocular mass of greater than 2 mm in thickness is seen on CT or MRI.

Introduction

Uveal melanoma is the most common primary intraocular malignancy and the second most common type of primary malignant melanoma in the body. The choroid is the most common site involved with 85%-90% of uveal melanomas arising from this region [1]. Uveal melanoma often presents with symptoms of decreased visual acuity, photopsias (flashes), floaters, and visual field defects [2]. Rarely, patients have no symptoms and uveal melanoma is detected on routine eye examinations. Choroidal melanoma is often diagnosed based on clinical examination by an ophthalmologist with experience in ocular oncology. It appears most frequently as a raised, pigmented subretinal lesion on ophthalmoscopic examination. Characteristic findings include thickness greater than 2 mm, associated serous retinal detachment, and intralesional lipofuscin [3]. Options for management include laser treatment, brachytherapy, proton beam radiotherapy, local resection, or eye enucleation. CT and MRI are not traditionally used for the diagnosis of choroidal melanoma though they may be useful for atypical lesions or where the diagnosis is in question. To our knowledge, there are no published cases of uveal melanoma noted incidentally on neuroimaging. Here we

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* Corresponding author. M. Tsukikawa.
E-mail address: tsukikaw@uw.edu (M. Tsukikawa).

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present 3 unusual cases in which choroidal melanomas were detected as incidental findings on neuroimaging for unrelated conditions in asymptomatic patients.

Case series

Case 1

A 77-year-old male underwent brain MRI due to several months of dysphagia. The scan showed an incidental finding of an ovoid T1 hyperintense lesion along the posterior superomedial aspect of the left globe (Figs. 1A and B). He had undergone cataract surgery in both eyes 4 years prior and at that time he was noted to have a non-concerning choroidal nevus in the superonasal periphery of his left eye (Fig. 1C). He denied any vision changes, flashes, floaters, or visual field defects.

The patient had a visual acuity of 20/20 in both eyes. Dilated funduscopic exam revealed an elevated, lightly pigmented choroidal lesion in the superonasal quadrant arising from the previously diagnosed benign nevus (Fig. 1D). Ocular ultrasound demonstrated a dome-shaped mass with acoustic hollowness, base 7.7 mm x 9.3 mm and thickness 3.6 mm.

Examination and imaging were consistent with a diagnosis of choroidal melanoma AJCC eighth Edition Stage IIA [4]. Systemic staging scans were unremarkable. The tumor was treated with proton beam radiotherapy. The patient maintained an expected post-radiation course. He developed radiation retinopathy and vitreous hemorrhage requiring vitrectomy surgery and laser delivery to the peripheral retina (Fig. 1E), which are common results of radiation for choroidal melanomas. At his last visit, 23 months since he completed PBRT, there was no evidence of local recurrence of the melanoma nor metastatic disease and best corrected visual acuity was 20/20 in both eyes.

Case 2

An 82-year-old male was hospitalized for cholecystitis and an episode of altered mental status. These findings prompted a CT head, which incidentally demonstrated a high density intraocular lesion in the posterior left globe. MRI brain was then obtained which redemonstrated a T1 hyperintense and T2 hypointense lesion (Figs. 2A-C). The patient had undergone cataract surgery in both eyes 4 years prior and no posterior lesion was noted at that time. Prior to the imaging finding, he had no symptoms in his left eye.

The patient had a best corrected visual acuity of 20/20 in the right eye and 20/30 in the left eye. Dilated funduscopic exam demonstrated a mushroom shaped lesion with associated serous retinal detachment in the inferonasal aspect of his left eye (Fig. 2D). Ocular ultrasound demonstrated acoustic hollowness with a basal dimension of 9.5 mm x 7.0 mm and thickness 10.4 mm.

Examination and imaging were consistent with a diagnosis of choroidal melanoma stage IIB [4]. Systemic staging scans showed no metastatic disease. Given the size and location of the tumor, and risk of potential complications and failure with radiotherapy, the eye was enucleated. Pathology demonstrated choroidal melanoma with mixed epithelioid and spindle cell morphology, stage pT3a [4] (Figs. 2E and F). Seven months after enucleation, the patient was doing well without signs of metastases.

Case 3

A 65 year old male was diagnosed with squamous cell carcinoma of the floor of the mouth and a pre-operative maxillofacial CT incidentally showed a mass in the posterior right globe measuring 1.4 cm in thickness (Figs. 3A and B). The patient reported weaker vision in the right eye since his youth and was told over 45 years ago that he had a “spot” in his right eye that was not growing. He had lost vision in that eye about 10 years prior to presentation which was attributed to a very dense cataract by his local optometrist. His medical history was unremarkable besides squamous cell carcinoma of the mouth.

The patient had no light perception in the right eye and a visual acuity of 20/20 in the left eye. Slit lamp examination of the right eye showed dilated conjunctival vessels, peripheral florid neovascularization of the cornea and iris, and an advanced brunescent cataract with no view to the retina. Ocular ultrasound revealed a dome-shaped mass, base 12.5 × 12.8 mm and thickness 10.8 mm.

Examination and imaging were consistent with a diagnosis of choroidal melanoma stage IIB [4]. Given the size and location of the tumor, and risk of potential complications with radiotherapy, enucleation was performed. Pathology demonstrated choroidal melanoma with mixed spindle and epithelioid morphology, stage pT3a [4]. Eight months after enucleation, the patient was doing well without signs of metastases.

Discussion

Uveal melanoma is the most common primary intraocular malignancy in adults and the second most common type of primary malignant melanoma in the body. The diagnosis of choroidal melanoma is based on clinical examination by an ophthalmologist with experience in ocular oncology. On the dilated funduscopic exam, choroidal melanoma appears most frequently as a raised, pigmented subretinal lesion in the posterior pole. Most choroidal melanomas are dome-shaped, but a collar-stud or mushroom configuration can arise after the tumor breaks through the choroid and begins to grow in the subretinal space. The more common differential diagnoses for choroidal masses include benign nevus, choroidal metastatic lesion, choroidal hemangioma, and choroidal granuloma.

The imaging modality most utilized in the diagnosis and management of suspected choroidal melanoma is ocular ultrasonography. Signs of choroidal melanoma include acoustic hollowness, choroidal excavation, and shadowing in the orbit. Ocular ultrasonography is also used for measurement of the tumor for radiation planning and identification of extrascleral extension. Other studies that can be utilized include fluorescein angiography, indocyanine green angiography, enhanced depth imaging optical coherence tomography, and autofluorescence photography. The Collaborative Ocular Melanoma Study reported a pathologically confirmed 99.5% diagnostic

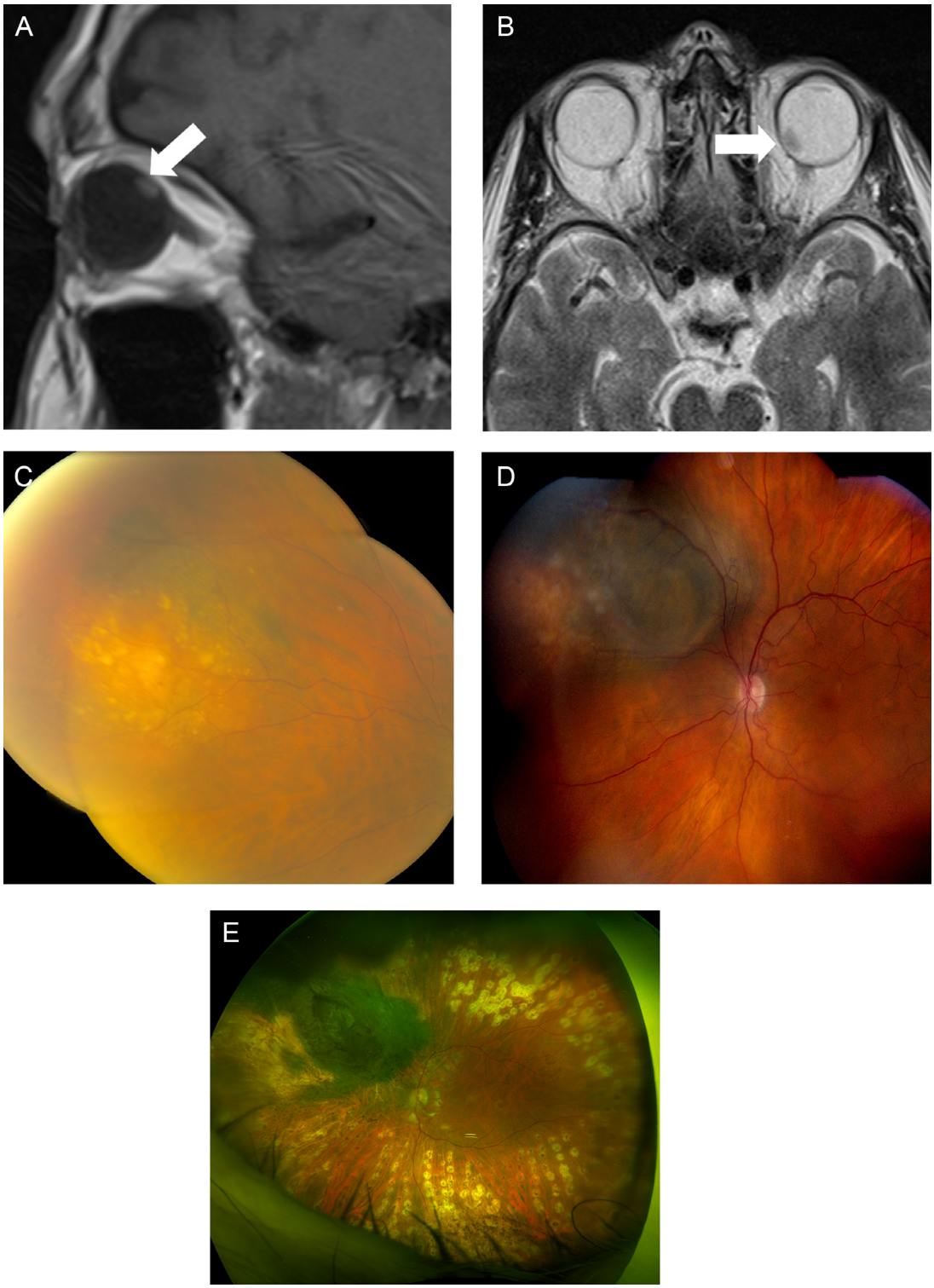

Fig. 1 – Patient 1. (A-B) Sagittal T1 weighted image (A) and axial T2 weighted image (B) from brain MRI for an unrelated condition demonstrated an incidental small T1 hyperintense and T2 hypointense mass centered at the superomedial aspect of the left posterior globe (arrows). There was no evidence of extraocular extension or optic nerve involvement. (C) Four years prior to presentation, a choroidal nevus was noted in the superonasal quadrant. (D) At presentation, an elevated, lightly pigmented choroidal lesion was identified in the same location. (E) Eighteen months after completing PBRT, he developed radiation retinopathy and vitreous hemorrhage requiring vitrectomy surgery and laser delivery to the peripheral retina.

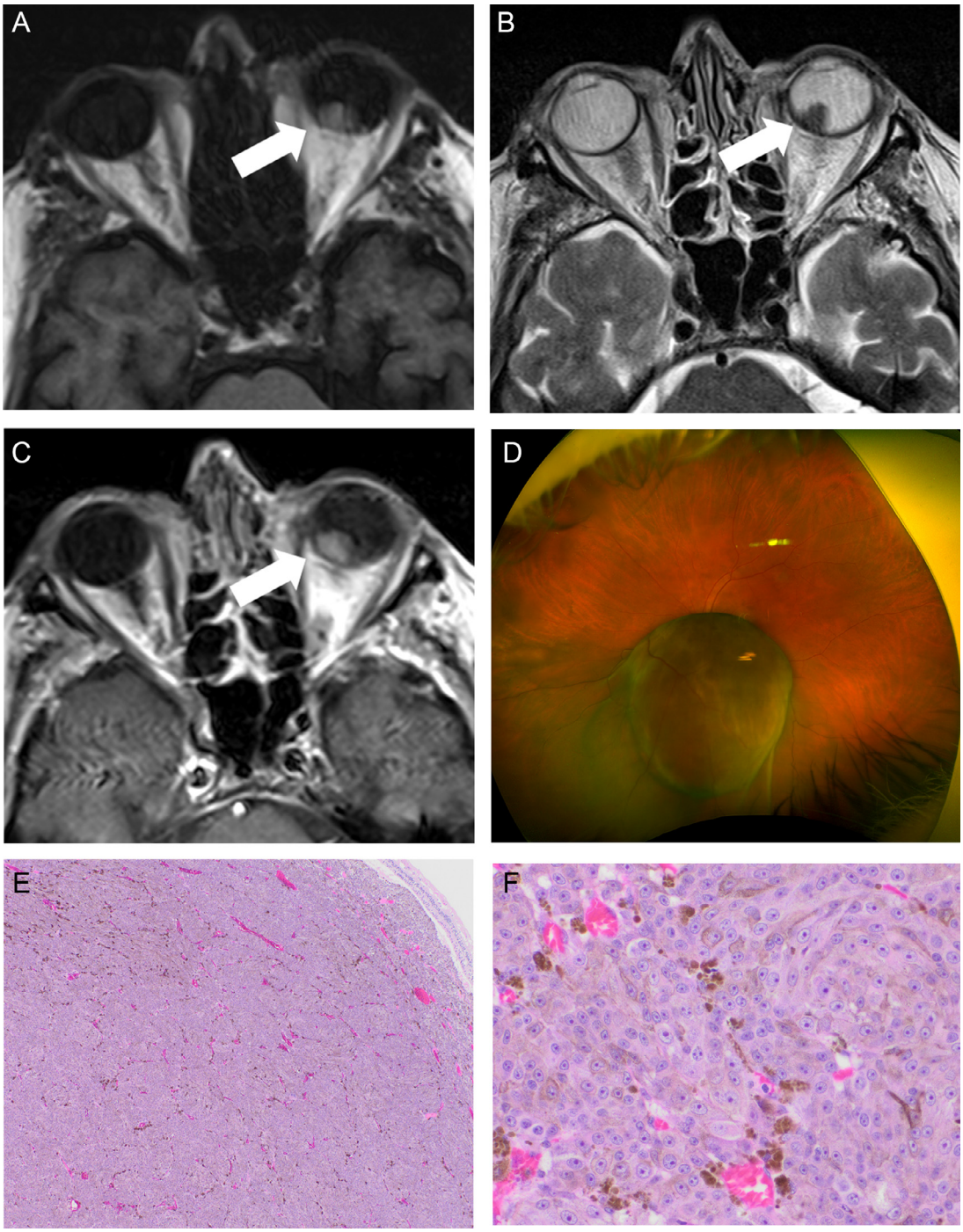

Fig. 2 – Patient 2. (A-C) Axial T1 weighted image (A), axial T2 weighted image (B), and contrast-enhanced axial T1 weighted image (C) demonstrated a small T1 hyperintense and T2 hypointense enhancing mass centered at the medial aspect of the left posterior globe (arrows). This mass was initially found on head CT which was performed due to altered mental status (not shown). (D) A mushroom-shaped lesion with associated serous retinal detachment was seen in the inferonasal quadrant. (E-F) The subretinal mass was found to be a pigmented, majority epithelioid, choroidal melanoma (H&E stains, original 40x and 400x).

accuracy for eyes diagnosed with these clinical signs and later enucleated [5].

CT and MRI are not routinely used for the diagnosis of choroidal melanoma. This is logical, as neuroimaging is not an important part of the diagnostic and referral patterns for uveal melanoma. In large studies, some including greater than 2000 patients, that have reported on patient presentation characteristics for nationwide cancer registries, none of the melanomas were found incidentally with neuroimaging [6,7].

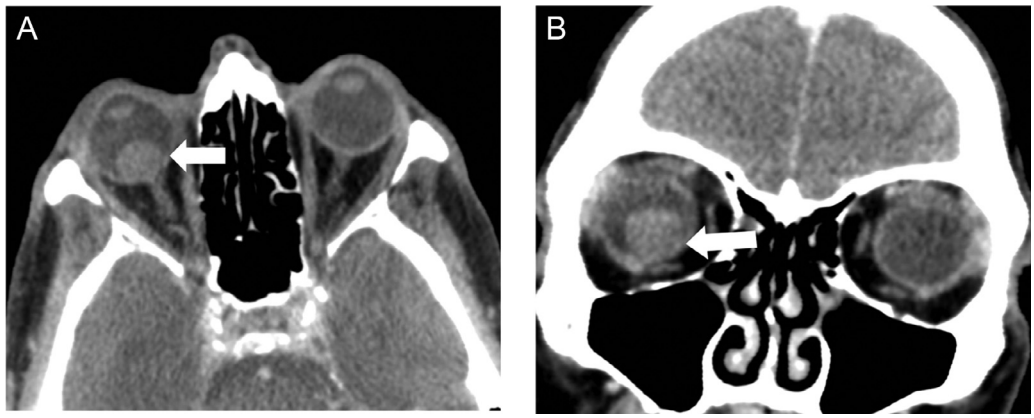

Fig. 3 – Patient 3. (A-B) Axial (A) and coronal reformatted (B) contrast-enhanced maxillofacial CT for an unrelated condition showed a mass centered in the inferomedial aspect of the right posterior globe (arrows), measuring 1.4 cm in greatest dimension.

There is a well-established role for MRI in the treatment of uveal melanoma, after it has been diagnosed. It has been used for guidance in both brachytherapy and proton beam radiation [8–10]. On MRI, choroidal melanoma typically appears hyperintense on T1 and hypointense on T2 with enhancement. Melanin has intrinsic T1 and T2 shortening effects, causing these T1 hyperintense and T2 hypointense characteristics. An approximate 25% of intraocular melanoma are amelanotic or present with low melanin content [11]. Tong et al. explained that even these pale lesions have varying T1 shorting effects based on melanin content and magnetic field strength utilized [12]. Other features such as size, location and enhancement characteristics would help to increase sensitivity of an intraocular neoplasm necessitating ophthalmology referral.

In this case series, choroidal melanoma was diagnosed in 3 patients due to incidental findings on neuroimaging. In all cases, the patients were asymptomatic and unaware of the growing melanoma. In each case, the melanoma was Stage II (1 patient with stage IIA, 2 patients with stage IIB) where the risk of subsequent metastatic disease is non-trivial. Documentation of the incidental finding on neuroimaging in all 3 patients led to prompt treatment, and all 3 patients remain metastasis free with a mean of 12.5 months of follow up. Correct diagnosis and referral of presumed uveal melanomas on orbital imaging can lead to earlier diagnosis, earlier disease stage at diagnosis, and therefore, an improvement in the expected mortality.

In conclusion, radiologists should maintain a high suspicion for uveal melanoma when an intraocular mass is seen on neuroimaging. On MRI, uveal melanoma typically appears hyperintense on T1 and hypointense on T2. A thickness of greater than 2 mm suggests a diagnosis of choroidal melanoma rather than choroidal nevus [3]. These patients should be referred to an ophthalmologist.

Patient consent

This research is part of a larger study entitled, “Retrospective study of ocular oncology treatments and outcomes.” This study has received approval from the University of Washington Institutional Review Board. Consent has been waived.

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