

Unsuspected and misdiagnosed posterior uveal melanoma following enucleation and evisceration in Ottawa-Gatineau

Seymour Brownstein, MD,^{*,†} André Jastrzebski, MD,^{*,†} Solin Saleh, MD,^{*,†}
David R. Jordan, MD,^{*} Steven M. Gilberg, MD,^{*} Brian C. Leonard, MD,^{*}
Bernard R. Hurley, MD^{*}

ABSTRACT •

Objective: There is a gap in the recent literature on the topic of clinically misdiagnosed and unsuspected posterior uveal melanomas (PUM) with a calculation of the frequency of these events for a specific geographical area. As the only ophthalmic pathology laboratory in our region, we determined the rate of these outcomes over a 16-year period.

Methods: We retrospectively reviewed 2558 consecutive ophthalmic pathologic specimens in the Ottawa–Gatineau region, of which 334 were eviscerations and 227 were enucleations. We calculated the frequency of clinically misdiagnosed PUM and of clinically unsuspected PUM. We also determined the rate of uveal melanoma undergoing enucleation.

Results: From 100 diagnoses of PUM, 2 (2.0%) cases were clinically unsuspected and 2 (2.0%) cases were clinically misdiagnosed. The rate of uveal melanoma undergoing enucleation was 5.6 cases per 1 000 000 of population per annum. From 2009 to 2012, the incidence of this event was 3.8 cases per 1 000 000 per annum, which was lower than the previous three 4-year increments.

Conclusions: We present the first and only single-centred, population-based data on the rates of unsuspected PUM and of clinical misdiagnosis of PUM in the era of modern diagnostic imaging. Our rate of clinical misdiagnosis is within the range of recent reports of this event. Unsuspected PUM occurred at a rate substantially lower than previously published. The incidence of uveal melanoma undergoing enucleation has decreased despite an increase in population, which reflects a shift in management from enucleation to radiation therapy.

Historically, clinically unsuspected posterior uveal melanomas (PUMs) were a common finding, with approximately 11% of all cases unsuspected clinically.^{1,2} Since 1980, we found 11 reported cases of unsuspected PUM after evisceration and 11 cases after enucleation in the English literature.^{3–11} A combination of increased awareness for unsuspected PUM and improved diagnostic imaging suggests that this is a rare finding, but others propose that these are highly underreported.⁸

Clinically misdiagnosed PUM was more commonly reported before 1974, occurring in 20% of cases.^{12,13} However, this rate declined steadily in subsequent studies,^{14,15} reaching a 0.48% clinical misdiagnosis rate in 1990 by the Collaborative Ocular Melanoma Study Group (COMSG),¹⁶ suggesting that the accuracy of the clinical diagnosis of PUM was no longer of concern. Nonetheless, clinically misdiagnosed cases continue to be reported, including 5 of 53 patients (9.4%) in one community center in the United States in 1997, indicating that this finding still is of concern.^{17–20}

We have a unique opportunity at our ophthalmic pathology laboratory, which is the only one serving all of the hospitals performing ocular surgery in the Ottawa–Gatineau region. By reviewing the records of our laboratory over a 16-year period, we have been able to calculate the incidence of PUM and the frequency of unsuspected

and misdiagnosed PUM in enucleated and eviscerated globes in our area.

METHODS

We performed a retrospective review of all cases processed by our laboratory from August 16, 1996, to August 15, 2012. We thus determined the average rate of occurrence of PUM for our region, which has an average population of 1 111 299 (obtained from Statistics Canada) over 16 years. Our study groups were all patients diagnosed unexpectedly with PUM after histopathologic examination of an enucleated or eviscerated globe and those who had been clinically misdiagnosed as having PUM. Four cases were identified.

We calculated the frequency of encountering a clinically unsuspected PUM, of clinically misdiagnosed lesions mimicking PUM, and the rate of PUM undergoing enucleation in our region, from the average annual population and the mean number of cases of PUM in our area over 16 years.

RESULTS

In our database, we found 334 eviscerations, 227 enucleations, and a total of 100 histopathological

Table 1—Incidence of unsuspected cases of PUM, cases misdiagnosed as PUM, number and incidence of PUM, and number of enucleations compared with eviscerations in the Ottawa–Gatineau region in 4-year increments between 1996 and 2012

	Aug. 16, 1996, to Aug. 15, 2000	Aug. 16, 2000, to Aug. 15, 2004	Aug. 16, 2004, to Aug. 15, 2008	Aug. 16, 2008, to Aug. 15, 2012	Total for 1996–2012
Unsuspected cases of PUM per 100 PUMs, n (%)	1 (1)	0	1 (1)	0	2 (2)
Misdiagnosed cases as a PUM per 100 PUMs, n (%)	1 (1)	0	0	1 (1)	2 (2)
PUM, n	32	22	27	19	100
PUM, n per 1 000 000 population per annum (average population)	8.0 (1 004 995)	5.1 (1 070 244)	6.0 (1 133 633)	3.8 (1 236 324)	5.6 (1 111 299)
Enucleations, n (%)	103 (70.1)	51 (34.2)	44 (29.5)	29 (25)	227 (40.5)
Eviscerations, n (%)	44 (29.9)	98 (65.8)	105 (70.5)	87 (75)	334 (59.5)

PUM, posterior uveal melanoma.

diagnoses of PUM in the 16-year period. Two of the 100 PUMs were clinically unsuspected (2.0%), and 2 additional cases were misdiagnoses (2.0%) (Table 1).

The incidence of PUM in enucleated and eviscerated globes in our region was 5.6 cases per 1 000 000 of the population per annum. Additionally, we calculated the rate of PUM in enucleated and eviscerated globes in 4-year increments between 1996 and 2012 and found that the incidence from 2009 to 2012 was 3.8 cases per 1 000 000 of the population per annum, which was lower than the previous three 4-year increments (Table 1).

Furthermore, we found a declining incidence of enucleation versus evisceration in the same quarterly timeline. In 1996–2000, 70.1% of treatments were enucleations, compared to a continuous decline to 25% treatment by enucleation in 2009–2012 (Table 1) ($p < 0.0001$ using the χ^2 test).

Report of cases

Case 1. A 55-year-old male presented with unilateral iritis, iris neovascularization, and a normal retinal examination in his right eye. He subsequently developed glaucoma refractory to medical management and underwent 7 surgical procedures, including trabeculectomies, a Molteno implant, and cyclophotocoagulation, after which he had no light perception vision, pain, and heterochromia in the right eye (Fig. 1A). He underwent a palliative enucleation in 1996. Four months later, he developed a subconjunctival nodule in the eye socket (Fig. 1B).

Histopathologic examination of the enucleated eye disclosed diffuse thickening of the ciliary body temporally and nasally (Fig. 1C) from an amelanotic ring melanoma composed of HMB45-positive anaplastic cells (Fig. 1D). Melanoma cells extended to the surgical margin along the limbal vessels and into the lumen of the Molteno implant (Fig. 1E, F), draining into the subconjunctival space. The subconjunctival nodule consisted of melanoma cells. The orbital contents were exenterated.

Three years later, solitary pulmonary and then cerebral metastases developed and were managed by surgical excision, radiation, and chemotherapy. Fourteen years

postenucleation, he developed disseminated metastases and was lost to further follow-up.

Case 2. An 86-year-old female with a prior macular hole repair was seen in 2008 with acute pain and light perception vision OS. Ocular examination revealed a massive vitreous hemorrhage, florid iris neovascularisation, and an intraocular pressure of 68 mm Hg. B-scan ultrasonograms disclosed appositional suprachoroidal detachments containing scattered dense opacities (Fig. 2A). Anti-vascular endothelial growth factor (bevacizumab) was injected into the eye, which initially resulted in improvement of the iris neovascularisation and intraocular pressure. However, 5 days later, intraocular pressure increased, along with worsening pain and chemosis. The left ocular contents were eviscerated for clinically diagnosed endophthalmitis in a blind, painful eye.

Histopathologic examination revealed mostly necrotic tissue containing melanin pigment and foci of epithelioid melanoma cells (Fig. 2B), which stained for HMB-45 (Fig. 2C). The diagnosis was a predominantly necrotic PUM.

Oncologic examination for metastatic disease was negative, and the patient was scheduled for enucleation. However, she died at home shortly thereafter of unknown causes, and no autopsy was performed.

Case 3. A 74-year-old male with a 2-month history of blurred vision in his right eye presented with a visual acuity of 20/400 and an enlarging inferotemporal amelanotic choroidal mass extending into the right macula (Fig. 3A, B).¹⁹ A-scan ultrasonograms revealed a lesion with medium internal reflectivity (Fig. 3C). The initial clinical examination was consistent with a metastatic lesion. Findings of repeated clinical, radiologic, and serological examinations were negative. An attempted fine-needle aspiration biopsy did not provide diagnostic tissue. Because of continued growth of the lesion, the eye was enucleated in 1997, approximately 1 year after initial evaluation, because of a concern that it harboured an amelanotic PUM. Gross dissection of the globe disclosed an amelanotic choroidal mass (Fig. 3D).

Histopathologic examination revealed a glandular tumour (Fig. 3E) that stained negative for carcinoembryonic

Download English Version:

<https://daneshyari.com/en/article/8791696>

Download Persian Version:

<https://daneshyari.com/article/8791696>

[Daneshyari.com](https://daneshyari.com)