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CASE REPORT Open Access

A case report: iridociliary melanocytoma associated with secondary glaucoma

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Abstract

Background Distinguishing between benign iridociliary melanocytoma and malignant melanoma presents a diagnostic challenge, particularly given the potential overlap in tumor growth patterns and clinical manifestations, especially when patients present with secondary glaucoma. Misdiagnosis may induce severe clinical consequences, including enucleation. Therefore, the judicious selection of biopsy or surgical techniques is crucial in both diagnosing and managing the condition.

Case presentation A 44-year-old female presented with uncontrolled elevated intraocular pressure (IOP) and a heavily pigmented iris lesion extending into the anterior chamber angle and adjacent ciliary body. Unexpectedly, standardized initial fine-needle aspiration biopsy (FNAB) yielded inconclusive results. Subsequent excisional surgery (partial iridocyclectomy and concurrent phacoemulsification) was performed to remove the tumor mass and treat cataract. Histopathological analysis confirmed the diagnosis as melanocytoma. Lens implantation followed upon normalization of IOP within 8 months. At the 2-year follow-up, the patient exhibited a satisfactory clinical outcome, with no tumor recurrence, achieving a best-corrected visual acuity of 20/40 and an intraocular pressure of 18.5 mmHg.

Conclusions This case underscores the importance of obtaining adequate tumor specimens for accurate diagnosis via FNAB in iris and ciliary body tumors. Additionally, for patients with secondary glaucoma, partial iridocyclectomy emerges as a promising intervention, addressing anterior chamber angle obstruction to alleviate IOP while facilitating histopathological diagnosis for subsequent management.

Keywords Iridociliary melanocytoma, Fine-needle aspiration biopsy, Secondary glaucoma, Iridocyclectomy

Background

Melanocytoma is a benign tumor characterized by dark pigmentation that can manifest in the optic nerve, choroid, iris, and ciliary body (CB) of the eye. An early study published in 1967 identified 2 melanocytomas among 89 CB lesions [1], while a recent Japanese study found 9 out of 32 eyes with ciliary body tumors [2]. Melanocytomas involving the iris and ciliary body are rare and can be easily misdiagnosed as malignant melanoma due to their large size, rapid growth, and significant pigment dispersion. Secondary glaucoma may develop due to trabecular meshwork blockage with pigment and obstruction of aqueous drainage caused by increased tumor mass in the anterior chamber angle. Treatment options depend on early diagnosis, with surgical intervention being necessary when glaucoma is present. Here, we present a case of primary iridociliary melanocytoma and discuss our insights into managing this condition.

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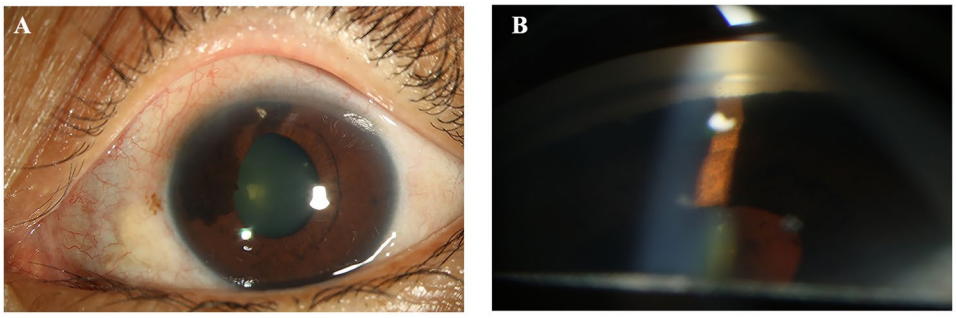

Fig. 1 Anterior segment photographs of the patient's left eye. **A**: Photograph showed a heavily pigmented and irregularly-shaped tumor lesion at the nasal iris from 8 to 1 o'clock. **B**: Gonioscopy examination showed the tumor involvement at the iris root and the pigment dispersion in the anterior chamber angle from 11 to 1 o'clock sites

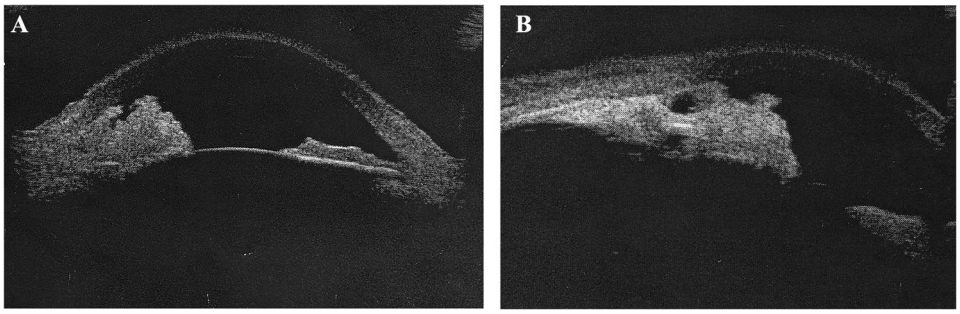

Fig. 2 Ultrasound biomicroscopy (UBM) examination of tumor lesion in the left eye. **A**: UBM revealed the non-flat, irregular mass at the iris and obstructs the adjacent chamber angle structure. **B**. The thicken and heightened reflectivity of the adjacent ciliary body suggested its involvement by the tumor lesion

Case presentation

A 44-year-old female patient was referred to our clinic due to recent-onset pain and vision impairment in her left eye spanning two weeks. She reported initial symptoms of decreased visual acuity, ocular redness, and discomfort localized to the left eye. Intraocular pressure (IOP) was 50 mmHg in the left eye and 16 mmHg in the right eye. Clinical examination identified a flat, pigmented lesion within the nasal iris. The patient had previously received a diagnosis of iris melanoma and secondary glaucoma at a local eye hospital, where enucleation surgery was recommended. Subsequently, she was referred to our institution for further assessment. Family and medical history were unremarkable.

A comprehensive examination, comprising ophthalmic assessment and systemic screening, was conducted. Best-corrected visual acuity (BCVA) was determined to be 20/166 in the left eye and 20/33 in the right eye. IOP was measured at 43 mmHg in the left eye and 16 mmHg in the right eye, without medication. Corneal opacity was observed, while the depth of the anterior chamber was within normal limits. A heavily pigmented lesion was identified in the nasal-superior iris, exhibiting a slightly elevated, diffuse, and ill-defined growth pattern. Few inflammatory cells and pigment dispersion were noted in the anterior chamber (Fig. 1A). No remarkable findings were observed in the right eye upon slit-lamp examination, except for mild cataract formation.

Treatment included rapid intravenous infusion of mannitol, along with topical administration of anti-glaucoma medications (carteolol & brinzolamide, twice daily) and tobramycin-dexamethasone drops (four times daily). Following these interventions, IOP decreased to 27 mmHg, and corneal clarity was restored within two days, prompting a gonioscopic examination. This examination revealed invasion of the angle by the tumor mass, resulting in heavy pigment deposition and obscuration of normal angle structures (Fig. 1B). Ultrasound biomicroscopy (UBM) depicted a thick, irregular iris mass with high reflectivity, extending from 8 to 12 o'clock and partially adhering to the corneal endothelium, with invasion into the adjacent ciliary body (Fig. 2). Fundus examination disclosed cup-to-disc ratios of 0.4 in the right eye and 0.6 in the left eye. Optic coherence tomography (OCT) identified peripapillary retinal nerve fiber layer defects in the superior, inferior, and nasal quadrants of the left eye. B-scan ultrasound examinations yielded unremarkable findings in both eyes. Regrettably, ultrasonography of the eyeball failed to identify the tumor.

A series of clinical laboratory examinations were conducted to investigate potential systemic disorders. Among these, only two tumor indicators, CYFRA21-1

(4.12 ng/ml) and Ferritin (189.10 ng/ml), were found to be slightly elevated. Computed tomography (CT) imaging of the chest revealed a few cord-like shadows in the lungs, suggestive of mild inflammation, with no suspicious tumor masses identified. Doppler ultrasound examinations of the entire abdomen and pelvis did not reveal any suspected tumors.

To ascertain the diagnosis of the lesion and guide subsequent treatment decisions, a fine needle aspiration biopsy (FNAB) was conducted. A trans-corneal FNAB procedure was employed, with the patient positioned supine and under topical anesthesia. A 26-gauge needle, connected to a 10-mL syringe via an extension tube (approximately 25 cm long), was utilized. The incision was made at the nasal aspect of the clear corneal limbus by the surgeon using the 26 G needle. The surgeon meticulously shook the needle within the tumor mass to facilitate the retrieval of additional tumor cells. Meanwhile, an assistant gradually withdrew the syringe plunger to aspirate tumor cells into the needle and tubing. Subsequently, the needle was removed from the eye and submerged into a cup containing sterile saline solution. The syringe plunger was then retracted to facilitate flushing of the biopsy specimen from the needle and tubing into the syringe. Balanced salt solutions were administered into the anterior chamber to maintain its depth as necessary. This procedure was repeated three times, and the syringe along with its contents were sent for pathological examination. The biopsy specimen was noted to contain brownish-black debris mixed with saline. Regrettably, the obtained samples were deemed insufficient for the definitive identification of the tumor cell type.

Surgery was scheduled for excision of the tumor mass for further pathological analysis upon obtaining the patient's consent. Following peribulbar anesthesia, a limbus-based conjunctival flap was created in the superior-nasal quadrant. Subsequently, the tumor, along with adjacent iris and ciliary body tissue, was excised through a corneoscleral tunnel incision. All suspicious tumor tissue was excised for histopathological examinations. Incisions were sutured, and a standard phacoemulsification procedure was performed. Intraocular lens implantation was deferred initially due to the uncertain prognosis.

A gross examination of hematoxylin and eosin (H&E) staining revealed heavily pigmented tissues, hindering the identification of characteristic tumor cells. Observations of bleached sections revealed two typical types of tumor cells, as reported before [3]: (1) Type I: large, polygonal, densely pigmented cells with giant melanosomes and abundant cytoplasm, small oval nuclei lacking prominent nucleoli, and (2) Type II: small spindle-shaped and sparingly pigmented cells with small melanosomes and few cytoplasm, elongated nuclei exhibiting prominent nucleoli. In this case, type 1 cells predominated among all observed tumor cells (Fig. 3). Immunohistochemistry (IHC) results indicated positivity for S-100, HMB45, and Melan-A (Fig. 4), while negativity was observed for P53. Ki-67-positive cells were few (less than 1%) and within the normal range. Based on these findings, the final diagnosis was concluded to be melanocytoma.

Postoperatively, the patient received Tobramycin-Dexamethasone four times daily and Tropicamide twice daily, along with 2% Carteolol and 1% Brinzolamide twice daily to manage IOP initially at 30–40 mmHg. Within eight months, IOP stabilized without medication, prompting IOP implantation to improve vision. The BCVA was 20/66 and the IOP was 20 mmHg on the first postoperative day. Tobramycin-Dexamethasone and Levofloxacin drops were prescribed for one month following surgery. Subsequent follow-ups (1, 2, 3, 6 and 12 months postoperatively) showed normal IOP. At the two-year mark, no tumor recurrence was observed (Fig. 5), with BCVA at 20/40 and IOP at 18.5 mmHg, yielding patient satisfaction.

Discuss and conclusion

Melanocytoma, initially described in 1962, denotes a benign tumor typically situated within the optic nerve head [4]. However, rare occurrences have been

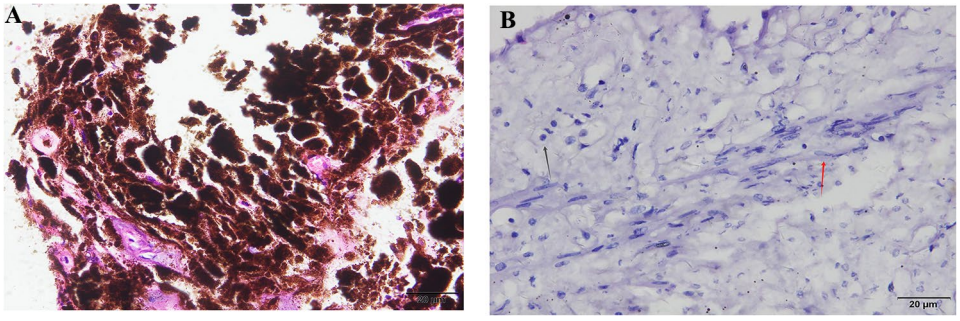

Fig. 3 Microscopic examination of the tumor specimen. (Haematoxylin and Eosin staining, H&E,×400). **A**: Micrograph showed the tumor cells with heavy cytoplasmic pigmentation. **B**: Bleach preparation showed large and polygonal cells with small nucleoli (Type I, black arrow) and spindle-shaped cells with round nuclei (Type II, red arrow)

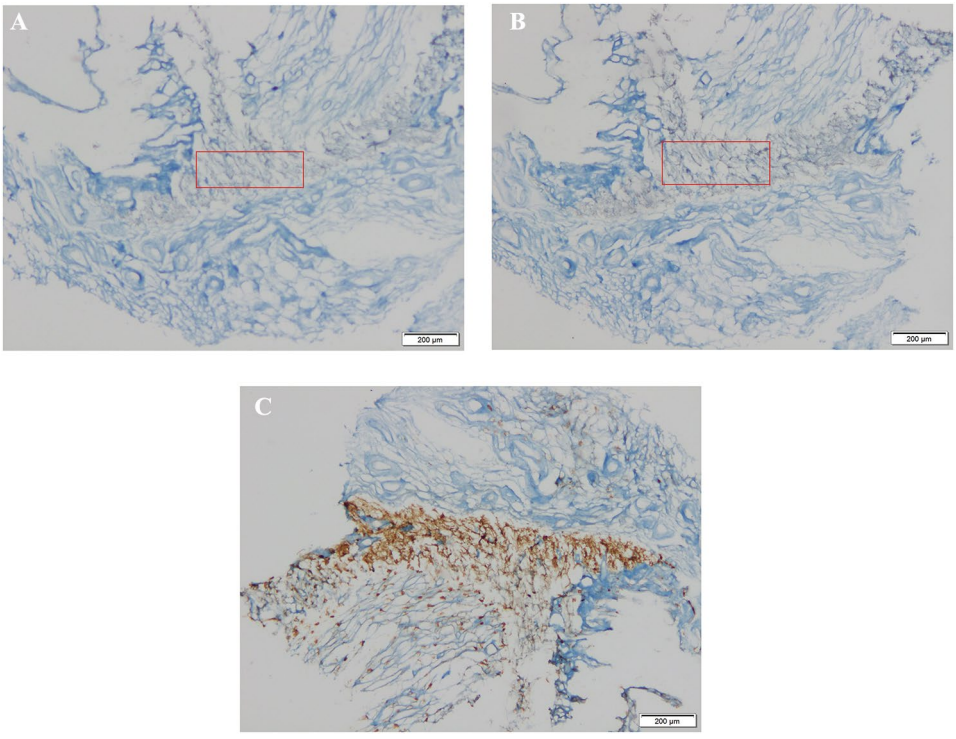

Fig. 4 Micrograph- IHC. **A** HMB45 staining positivity(×100): The red box area contains representative positive tumor cells, with HMB45 positive cytoplasmic staining. **B** MELANA staining positivity(×100): The red box area contains representative positive tumor cells, with MELANA positive cytoplasmic staining. **C** S-100 staining positivity(×100):The brown-stained area represents tumor cells, with positive S-100 staining observed in both the cytoplasm and nucleus

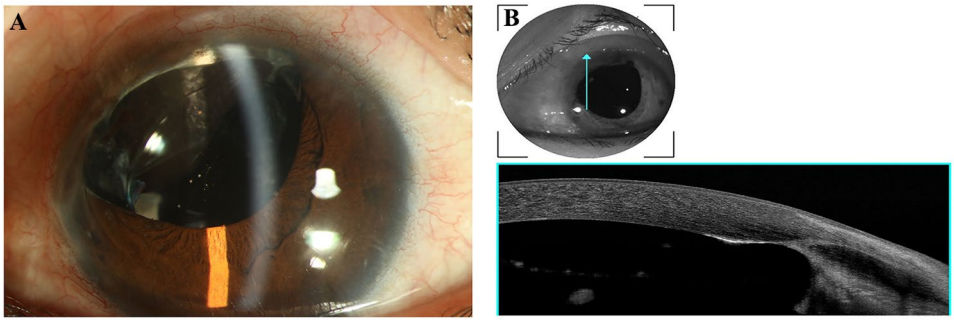

Fig. 5 Anterior segment photograph of the patient's left eye at postoperative 2 years visits. **A**: Photograph showed no tumor recurrence, residual pigment keratic precipitates in the previous surgical sites. **B**: High-resolution image of the anterior-segment optical coherence tomography showed no tumor recurrence in the iris and adjacent ciliary body

documented originating from various ocular structures including the iris, ciliary body, choroid, sclera, and conjunctiva [4]. Iris lesions commonly present as flat, well-defined growths localized to the chamber angle and usually remain stable over time. In contrast, occasional cases display diffuse tumor growth with irregular surfaces and pigment dispersion, posing a significant risk for secondary glaucoma, as observed in our patient [5].

Distinguishing melanocytoma from melanoma remains a challenge. While melanocytoma can resemble iris nevus, rare cases show benign lesions can develop into melanoma [6, 7]. Manifestations in our case including irregular tumor surface, heavily pigment dispersion, and secondary glaucoma were considered as the possible indicators of melanoma [7].

Ultrasonography such as A-scan, shows inconsistent findings regarding melanocytoma characterization [4, 8]. Although a prior investigation highlighted the potential of high-resolution B-scan in conjunction with A-scan in the identification and monitoring of patients with melanocytoma, ultrasound remains incapable of distinguishing between melanocytoma and melanoma [9]. In our case, we suspected that the failure of ocular ultrasonography to detect the lesion could be attributed

to subtle morphological changes involving the iris and ciliary body. On the other hand, UBM emerges as a preferable approach for evaluating such lesions, providing crucial details regarding growth patterns, tumor thickness, angle adhesion, and ciliary body involvement [10]. In our case, UBM clearly showed the irregular surface and thickening of the lesion of iris and the adjacent ciliary body with medium to high reflectivity, indicating ciliary body involvement. Since melanocytomas of the iris and ciliary body exhibit a variety of internal reflectivity patterns, as demonstrated by both our case and previous cases [11, 12], we suggest that it remains challenging to determine the malignancy of the tumor solely based on its internal reflectivity on UBM [13].

Ocular melanocytomas typically exhibit hyperintensity on T1-weighted and hypointensity on T2-weighted magnetic resonance imaging (MRI) scans, similar to melanoma [4, 14, 15]. However, distinguishing between the two based on MRI findings alone is difficult [16]. It is worth mentioning that (123) I-IMP SPECT has emerged as a promising tool for diagnosing malignant uveal melanoma, providing an option of distinguishing melanoma from melanocytoma [17]. However, in our case, neither MRI nor (123) I-IMP SPECT imaging was performed.

Biopsy plays a crucial role in diagnosing iris and ciliary body tumors like melanocytoma, as it provides cells and tissues for cytologic and histopathologic examination. Methods range from excisional techniques (such as iridectomy and iridocyclectomy) to less invasive approaches like FNAB, vitrector-assisted biopsy, and Kelly punch-assisted biopsy.

FNAB, though widely used with rare reported complications [18–20], may provide limited diagnostic accuracy due to cell quantity. Despite Shields and associates reported FNAB is effective in diagnose iris tumor with high sensitivity in 1993 [19] and 2006 [20], they emphasize the success depends on the experience of surgeon and cytopathologist. In our case, cytological examination followed FNAB was inconclusive for excluding melanoma. This highlights the importance of obtaining adequate tumor tissues for diagnosis. Finger et al. [10] proposed an aspiration-cutter-assisted biopsy through the iris root, which offers both cells and tissue fragments for diagnosis and may provide greater diagnostic insight compared to FNAB.

Management of melanocytoma typically involves observation, with surgical options including iridectomy, iridocyclectomy, or enucleation due to melanoma concerns [6, 7]. Considering the failure of FNAB to rule out melanoma, and the necessity to treat glaucoma [6, 7], a sector iridocyclectomy was performed for both diagnostic and therapeutic purposes. Histopathological analysis of the excised tissue revealed the presence of two distinct types of melanocytoma cells, with a predominance of type 1 cells over type 2 cells, and no mitotic activity was observed. Consistent with previous literature, small spindle cells were predominantly localized at the periphery of the tumor [21]. The histopathological evaluation definitively confirmed the diagnosis as melanocytoma.

In contrast to precedent cases [6], our patient exhibited transiently elevated IOP within the first postoperative month, which was effectively managed with a single anti-glaucoma eye drop. Subsequently, IOP normalization occurred within 8 months without further medication. The identified risk factors for glaucoma in this condition included tumor invasion into the chamber angle and pigment dispersion [22]. Notably, residual pigment keratic precipitates were discerned in the surgical site at the 2-year follow-up, suggesting that the primary mechanism of glaucoma in this instance may be attributable to tumor-induced trabecular outflow obstruction and consequent angle closure.

In this case, the eyeball was saved from enucleation and resulting in satisfied BCVA and IOP. As a result, the patient has experienced a successful clinical outcome, and is currently undergoing follow-up care. Reflecting on this diagnostic and therapeutic journey, we underscore the importance of tumor biopsy, we advocate for excisional procedures in patients presenting with secondary glaucoma, as these interventions not only supply adequate tissue for accurate diagnosis but also address chamber angle obstructions, thereby facilitating IOP reduction.

Abbreviations

CB	Ciliary body
IOP	Intraocular pressure
BCVA	Best-corrected visual acuity
FNAB	Fine needle aspiration biopsy
UBM	Ultrasound biomicroscopy
OCT	Optic coherence tomography
CT	Computed tomography
H&E	Hematoxylin and eosin
MRI	Magnetic resonance imaging

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Author contributions
YS and JG conceptualized and designed the study, YS drafted the initial manuscript, JG and ZFW reviewed and revised the manuscript. JZ and JG followed the patient and managed the treatment. YYC performed the histopathologic examination and gave advise to the manuscript.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethical approval
This study was conducted with approval from the Ethics Committee of the Third People's Hospital of Chengdu (2024-S-155).

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Competing interests
The authors declare no competing interests.

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