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Iris Melanocytoma in a Child: Clinical and Histopathological Findings

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Conflict of interest: None declared

Patient: Male, 3-year-old
Final Diagnosis: Iris melanocytoma
Symptoms: Eye pain and redness • photophobia
Medication: —
Clinical Procedure: —
Specialty: Ophthalmology

Objective: Rare disease

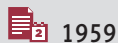
Background: Melanocytoma is rare and can affect any part of the uveal tract. In rare cases, iris melanocytoma shows signs of growth, with extrascleral extension that mimics melanoma. This phenomenon makes clinical differentiation between the 2 pathologies particularly challenging.

Case Report: A 3-year-old boy presented with recurrent ocular inflammation. Examination revealed a large, solid, homogeneous mass in the inferior quadrants of the iris, with secondary localized corneal edema. The lesion did not extend to the ciliary body and fundus examination showed no lesions in the posterior segment, including the head of the optic nerve. The patient underwent a sectoral iridocyclectomy and excisional biopsy of the lesion in the iris. Histopathology of the lesion confirmed the diagnosis of iris melanocytoma.

Conclusions: The differential diagnosis for a mass in the iris is broad, ranging from benign cysts to melanoma, which is a life-threatening ocular condition. An iris melanocytoma always should be considered in the differential of these masses, despite their exceedingly low incidence. Although iris melanocytoma mainly manifests in patients who are middle-aged or older, it should be suspected in young children, as underscored by the present report.

Keywords: Iris • Iris Diseases • Iris Neoplasms

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Background

The term “melanocytoma” has been used primarily to describe a deeply pigmented mass on the head of the optic nerve [1]. Less commonly, melanocytoma can affect any part of the uveal tract, as has been documented, and it has been confirmed histopathologically to be totally distinct from malignant melanoma of the iris [2]. Although melanocytoma is considered to be benign, unlike melanoma of the iris, its secondary effects are consequential. Tumor necrosis with resultant shedding, which leads to anterior chamber seeding, iris stromal seeding, ectropion uveae, and secondary glaucoma, is a well-known effect of the mass lesion [3]. In addition, in rare cases, iris melanocytomas show signs of growth with extrascleral extension that mimics melanoma. This phenomenon makes clinical differentiation between the 2 pathologies particularly challenging [4]. Progression of benign melanocytoma to malignant melanoma has been reported [5]. Given these factors, histopathological examination of the specimen is the criterion standard for diagnosis of iris melanocytoma. Here, we report the case of a child with a confirmed diagnosis of iris melanocytoma after excisional biopsy.

The present case report complies with the principles of the Declaration of Helsinki. Oral but not written consent to publish the case was obtained. Our report contains no personal information that could lead to identification of the patient

Case Report

A 3-year-old boy with no significant medical history presented to the Emergency Room of King Khaled Eye Specialist Hospital with a 3-month history of photophobia of the right eye, and swelling of the upper lid of that eye. During that period, he had experienced intermittent redness in the same eye, which was partially relieved by a topical steroid that had been prescribed for him at another facility. Eight months before, the patient had experienced blunt trauma to the same eye, in the form of a blow from a fist. He had been born at full term, by spontaneous vaginal delivery, and his mother had no known prenatal infections.

On ophthalmic examination, the patient’s visual acuity was fix and follow in each eye. His intraocular pressures (IOPs) were 13 mmHg in the right eye and 18 mmHg in the left.

Biomicroscopic examination of the left eye was unremarkable. Examination of the right eye showed an intact globe. The conjunctiva was chemotic, mainly inferiorly and temporally. The cornea showed some dispersed pigmented keratic precipitates inferiorly with localized inferior corneal edema. The pigment cells in the anterior chamber had an aqueous flare reaction

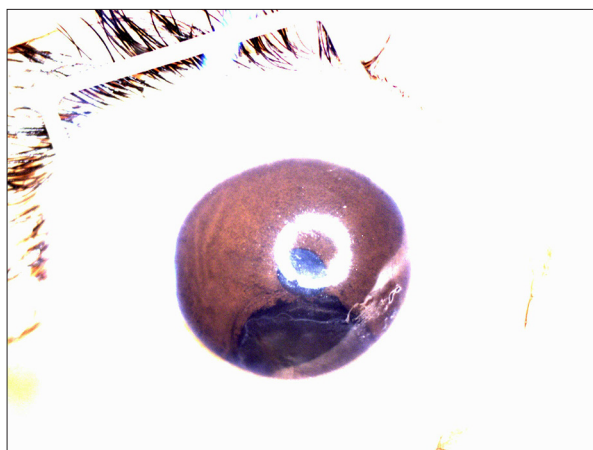


Figure 1. Image of a large, jet-black lesion in the iris in close proximity to the pupillary margin with mild corneal edema atop it.

that was almost grade 1+, but no white blood cells were detected. In addition, the chamber was noted to be formed, except inferotemporally, where a jet-black pigmented lesion was visible in the inferior quadrant of the iris between the 5 and 7 o’clock positions. It appeared to be solid and was close to but did not involve the pupil. The peripheral part of the lesion invaded the angle (Figure 1). Extension of the lesion obstructed the gonioscopic view into the chamber angle between 5 and 7 o’clock. The remainder of the angle was open, with a moderate amount of pigmentation covering the scleral spur and trabecular meshwork (Figure 1). There were anterior capsular changes, which manifested as mild lens opacity near the lesion. No intrinsic vascularity was seen within the mass nor was there neovascularization in the other part of the iris. Posterior segment examination of both eyes was unremarkable.

Because the lesion was obscuring the angle, ultrasound biomicroscopy (UBM) was used to evaluate the depth of penetration into the iris and surrounding tissues. The mass in the iris was found to be solid with no calcifications. It was closing the angle inferotemporally but the ciliary body was spared from involvement (Figure 2A). On UBM, a broad adhesion was clearly visible between the corneal endothelium and the mass in the iris (Figure 2B). An amplitude scan showed high internal reflectivity (Figure 2C). Given the constellation of findings, the decision was made to perform a surgical excisional biopsy during sectoral iridocyclectomy, which involves removing parts of both the iris and ciliary body. Iridocyclectomy was chosen instead of iridectomy alone because while imaging showed that the ciliary body was not involved, there was suspicion about microscopic seeding into the ciliary body, which would not be detectable on UBM and could induce recurrent inflammation postoperatively if left untreated. A conjunctival peritomy was performed adjacent to the involved area by creating a superficial scleral flap that was 3 to 4 mm larger than

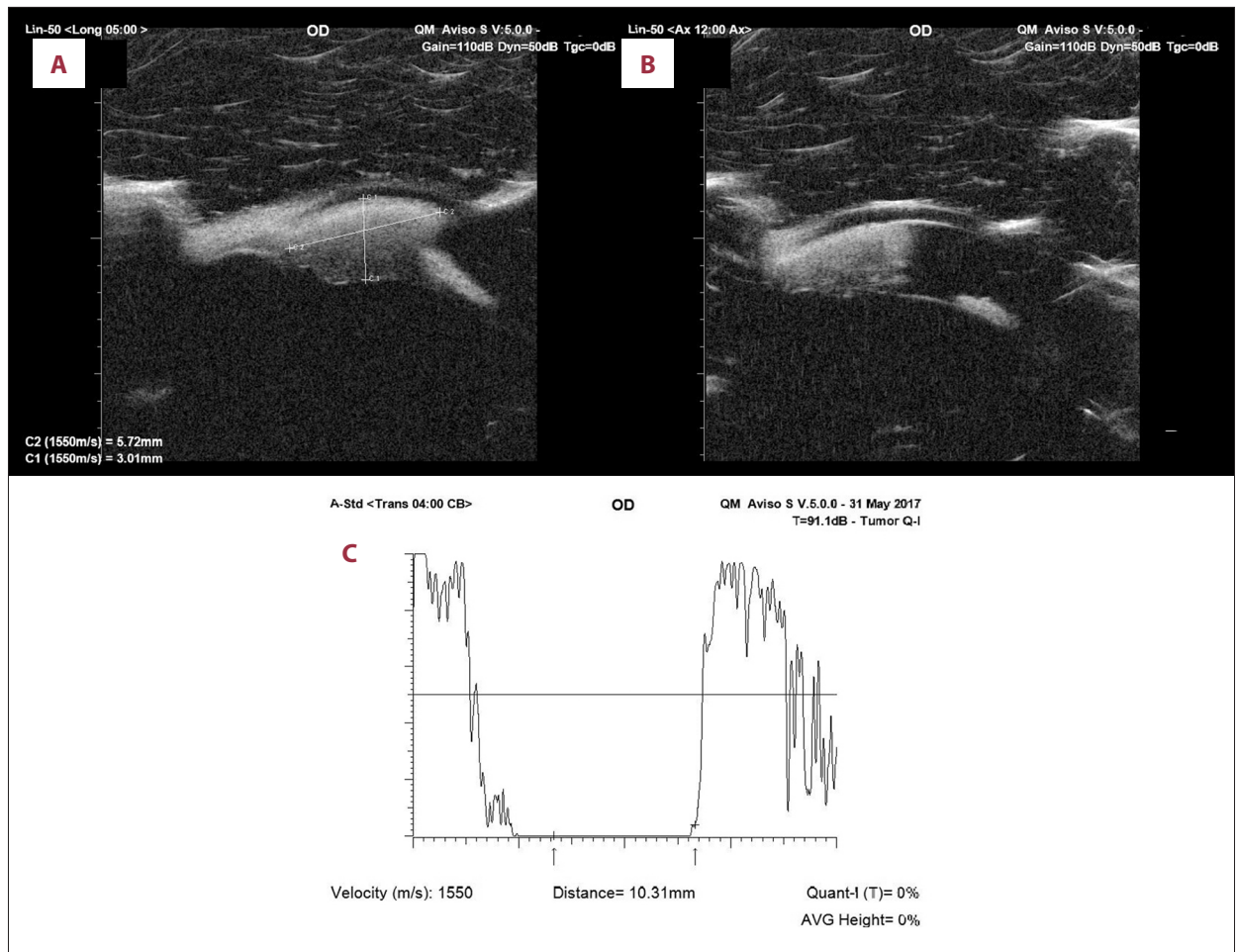


Figure 2. (A) Ultrasound biomicroscopy (UBM) shows a solid iris mass that measures 3.01 mm in depth and 5.72 mm in width with no calcification or ciliary body involvement. (B) UBM showing the attachment between the iris mass and the corneal endothelium. (C) An amplitude scan shows the highly reflective mass.

the tumor. Entry into the anterior chamber was achieved by creating a deep scleral flap the same size as the tumor and extending it into the limbus. Viscoelastic was injected to stabilize the anterior chamber, tamponade the corneal endothelium, and free the posterior synechiae, which had been observed during surgery before the iris and ciliary body were cut with intraocular scissors to create a 1-mm clear margin. The lesion was removed through the scleral incision and the specimen was sent for histopathologic analysis. Large polyhedral nevus cells were seen, which contained a large amount of melanin and obscured the nuclear details (Figure 3), confirming the diagnosis of melanocytoma.

On the day after surgery, the patient received topical atropine 1%, prednisolone acetate 1%, and moxifloxacin eye drops. He was stable with no pain or other symptoms. Examination showed mild conjunctival chemosis and a clear cornea except inferiorly. In that area, there was haze in addition to brown pigmentation at the site of the prior adhesion, the corneal suture

was in place with no leak or defect, and the anterior chamber was deep with no cells or flare. There was an inferior defect in the iris after excision and anteriorly, there was capsule pigmentation from previous synechiae. An early cataract was noted and examination of the retina showed a healthy disc and flat retina with no breaks or holes. The patient's IOP was normal.

During the first visit after surgery, the patient's parents reported that he was having no issues and was sleeping well, with no pain. Examination findings were similar to those documented immediately after the procedure (Figure 4A, 4B). On further follow-up, the patient continued to have stable vision with fix and follow in both eyes, normal IOP, and no recurrence of inflammation or new symptoms. The cataract that was seen previously showed no signs of progressing or involving the visual axis. The patient was referred to Pediatric Ophthalmology for cycloplegic refraction and management of amblyopia.

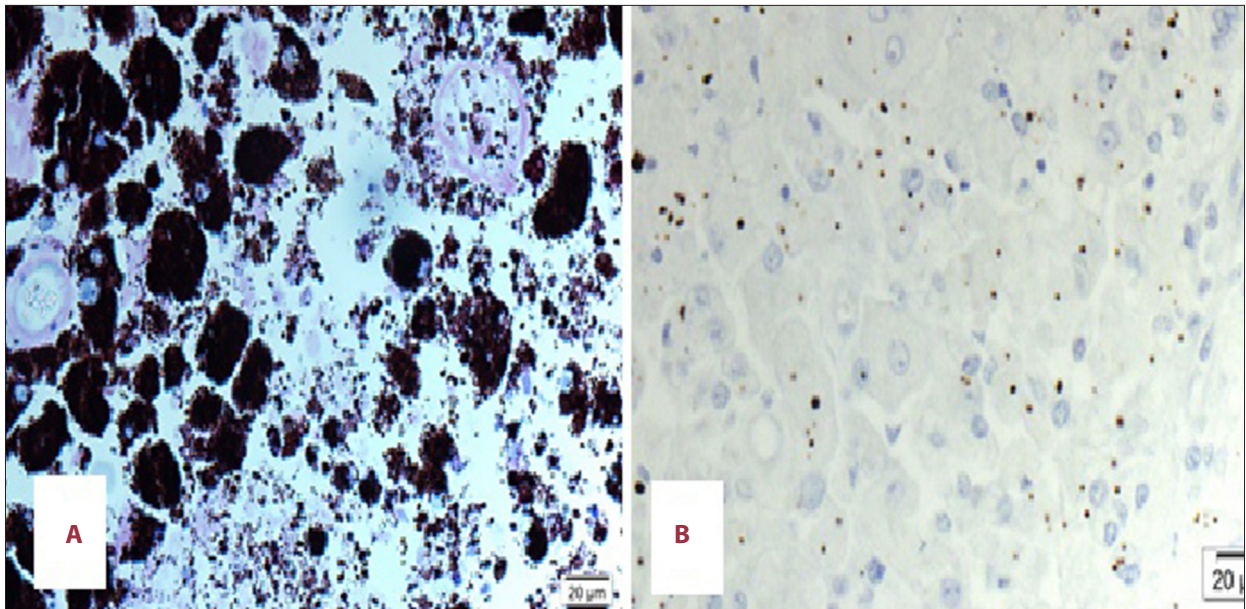


Figure 3. (A) Histology shows that the tissue of the iris is infiltrated by large polyhedral nevus cells with dense pigmentation and obscure cellular details (hematoxylin and eosin 400×). (B) Large bland cells with oval to round nuclei and indistinct nucleoli with abundant cytoplasm, consistent with the diagnosis of melanocytoma (melanin A bleached 400×).

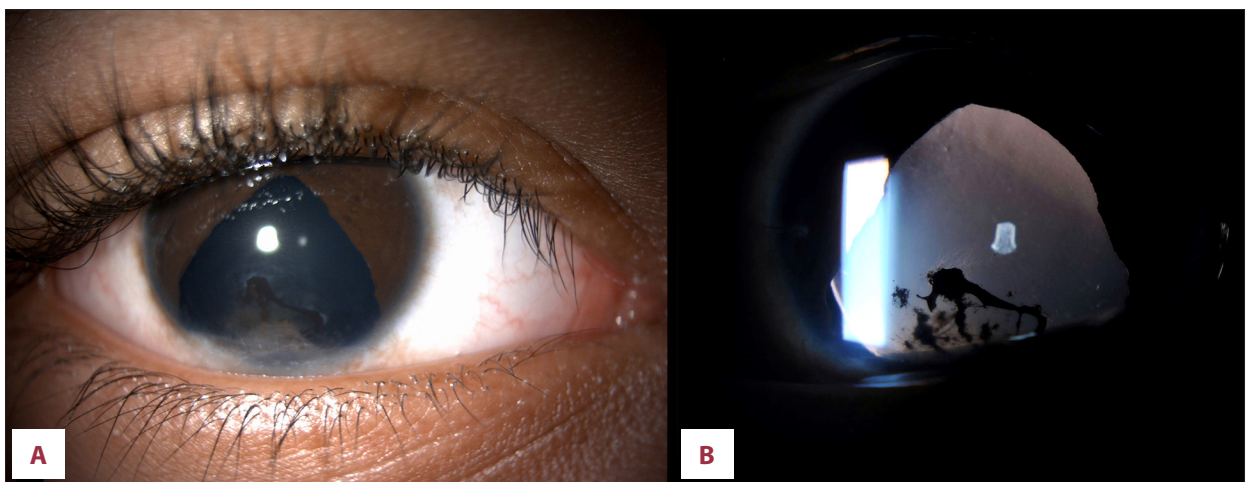


Figure 4. (A) Slit lamp examination after excision shows inferior corneal haze with inferior iris defect and pigmentation of the cornea due to prior adhesion. (B) A retroillumination technique highlights the area of corneal pigmentation.

Discussion

Iris melanocytoma is a rare cause of localized or diffuse iris masses and always should be considered in differential diagnosis of such lesions [2,6]. It is a distinctive iris nevus because of clinical and histopathological characteristics that distinguish it from other iris masses [3]. The rarity of iris melanocytoma is underscored by a report by Shields et al, who found it in only a single eye among 200 (0.6%) in patients referred for iris masses. That is an incredibly low incidence compared to other iris lesions that simulate iris melanoma (pseudomelanomas), such as iris nevi (31%) and cysts (38%) [6].

Adults typically are affected by melanocytoma and it is very rare in patients younger than age 10 years. Green et al reported a case in a 6-year-old and Shields et al reported another case in a 9-year-old patient [4,7]. The mean age of patients in the largest study of 47 eyes with iris melanocytomas was 36 years and only 4 patients were younger than age 10 years [3]. Patients in that study ranged in age from 3 to 67 years, whereas our patient was the same age as the youngest individual in the largest study of iris melanocytoma. This fact underscores the importance of always considering iris melanocytoma in patients of any age who present with an iris mass, despite the fact that it is rare and typically manifests in adults.

Iris melanocytomas can occur in any quadrant of the iris or it can present diffusely, but the inferior quadrant is the one most commonly involved [3,8]. This could be a helpful clue in aiding in diagnosis of these lesions. Although melanocytoma is considered benign, secondary consequences of these lesions have been well documented. The effects are associated with either necrotic shedding of the mass or, in rarer cases, growth that occurs in a minority of patients and mimics melanoma or may indicate malignant transformation [4,5]. The mechanism of mass necrosis that results in shedding is not fully understood but has been hypothesized to be related to insufficient circulation in a highly metabolic mass [9]. Although the majority of melanocytomas show no signs of growth, it has been reported in 23% of cases at 5 years and 48% of cases at 10 years [10]. The growth can compromise visual function through blockage or development of a secondary cataract when the lesion comes into contact with crystalline lens, as occurred in our patient. The propensity for shedding is the factor responsible for the variability of presentations, which include iris or anterior chamber seeding leading to ocular inflammation, seeding into the angle that can induce secondary glaucoma, and even iris heterochromia, especially in diffuse lesions [5,8].

Our patient had a history of trauma, but he presented with persistent ocular discomfort and recurrent inflammation that was not associated with the trauma. The best explanation for his symptoms were the findings from the ocular examination, which showed mild anterior chamber inflammation that resembled the effects of lesion shedding. Another interesting finding we noted was localized corneal edema over the lesion due to corneal compression by the massive melanocytoma, which resolved after excision. This secondary effect and finding have not been previously reported in the literature to be associated with iris melanocytoma.

Different modalities have been suggested for diagnosis and treatment of iris melanocytoma. Clinical observation is appropriate for stable lesions that show no sign of malignancy on presentation or during follow-up [3]. For unstable lesions that show progressive growth or when the diagnosis is questionable, many methods have been used, including fine-needle aspiration (FNA) or, more commonly, excisional biopsy with

surgical iridectomy, iridocyclectomy, or iridogoniocyclectomy, depending on which part of the iris is involved [4,5,11]. FNA reportedly is highly effective for diagnosing these lesions, with accuracy reaching 99%, and it is less invasive than excisional biopsy. The major disadvantages include intralesional hemorrhage resulting in hyphema or spread of the lesion after aspiration [11]. Excisional biopsy can be used to diagnose and manage the lesion with debulking, preventing future adverse events associated with lesion shedding [3].

Because our patient was a child with a questionable diagnosis and a very unstable lesion, en-bloc excision of the whole lesion was performed. FNA was not feasible, given the risk of hyphema after aspiration, which could have affected the child's visual acuity, especially if coupled with corneal staining during development of his vision. The patient's biopsy confirmed the diagnosis of iris melanocytoma despite a clinical scenario that suggested a post-traumatic cyst of the iris. The patient had characteristic large polyhedral cells that contained a copious amount of melanin pigment, which explains the jet-black color that manifested during presentation. The absence of atypia on histopathology slides and of either spindle or epithelioid cells, which are characteristic of malignant melanoma of the iris, are the only way to confidently distinguish between a benign mass and a malignant tumor [2,6]. Therefore, clinical interpretation of masses in the iris always should be supported by results of pathological analysis.

Conclusions

Iris melanocytoma is very rare and always should be suspected in any patient who has a mass in the iris, even a child. It has very subtle findings that can differentiate it from other iris masses, including malignant melanoma of the iris. Excisional biopsy to confirm and debulk the lesion is the criterion standard approach in such cases, especially when the diagnosis is questionable or the lesion is unstable.

Conflict of Interest

None.

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