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IRIS MELANOMA: A CASE REPORT

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SUMMARY – Although its location makes the iris easily accessible for examination, differentiation of melanocytic malformations such as nevi or melanomas is difficult. Inaccurate judgment may lead to morbid ocular and systemic complications. If tumor is detected in early phase, prognosis is good. Management decisions for iris melanoma depend on clinical features. In most cases, local resection with careful assessment for iridocorneal angle involvement achieves long term tumor clearance with an acceptable morbidity. We report on a 52-year-old man who presented to an eye clinic with a pigmented lesion on his right eye iris. After thorough local and systemic examination, and three-year follow up, we performed iridectomy with sclerotomy and iris reconstruction to reduce postoperative photophobia. The patient was satisfied with functional and cosmetic result. No further treatment was indicated.

Key words: *iris melanoma, iridotomy*

Introduction

An iris nevus is a cluster of abnormal neuroectodermal melanocytes in the iris stroma[1]. Most iris nevi tend to remain dormant throughout life and do not require any treatment. Rarely, iris nevus transforms itself into a malignant melanoma. Differentiating benign iris nevi from iris melanoma can be difficult. The usual symptom of iris melanoma is a visible spot on the iris or discoloration of the iris in one eye. Many patients with iris melanoma have no symptoms, and the lesion in these patients usually is detected on routine eye examination. If the tumor begins to grow or if there are symptoms, an operation is needed.

A generally more benign prognosis and historically high false positive diagnosis rate have led to a trend in recent years towards conservative management of suspect melanocytic iris tumors. This involves periodic observation of the lesions and interventions aimed at preserving the eye and vision if there is evidence of more aggressive behavior; enucleation now being reserved for large unresectable tumors[2]. The prognosis is generally good. About 5% develop metastases. The 5-year mortality rate is *2%-3%*.

Mixed or epitheloid histology is more likely in the presence of two or more of the features of malignancy and may justify earlier intervention.

On resecting iris melanoma, careful assessment for iridocorneal angle involvement is important in treatment planning. Iris reconstruction has a useful role in reducing postoperative photophobia.

Melanoma of the iris is rare. It makes about 5% of uveal melanomas (the proportion varies between 3% and 12%). The annual incidence is estimated to vary between 0.2 and 0.9 *per* million population. It is about 3 times more common in patients with light (blue/grey) than brown irides. The tumor is almost always unilateral and arises from a pre-existing nevus. Conditions associated with or predisposed to early onset of uveal melanoma are ocular melanocytosis, nevus of Ota, dysplastic cutaneous nevi, familial melanoma and neurofibromatosis-1. Sun exposure is still a questionable risk factor. The great majority of iris melanomas occur on the inferior half of the iris, which further implicates sun exposure.

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Case Report

A 52-year-old man came to an eye clinic because he noticed a lesion on his right eye iris. He had no pain, no visual loss or any other symptom. The slit lamp examination showed a dark pigmented nodule of the right eye iris which was of a small seed size. It was located in the inferior temporal quarter. We noticed no surface vascularity of the lesion or conjunctival sentinel vessel. There was a discrete localized opacity on the anterior capsule of the lens. Nothing abnormal was found in the left eye. We performed orbital ultrasonography and magnetic resonance imaging (MRI), and found no signs of tumor spread. General examination was performed too. There was no extraocular pathology.

According to algorithms we decided to follow up lesion with documentation. The patient was observed every six months (slit lamp examination, gonioscopy, photography, ultrasonography, orbital MRI, general examination). After three years we noticed first signs of progression. The lesion was highly pigmented, 3x3 mm in size, slightly more prominent and with surface vascularity present. There was no cataract progression and no signs of ciliary body affection.

We performed iridectomy with sclerotomy and iris reconstruction to reduce postoperative photophobia. The operation was performed in local anesthesia. Histopathologic examination confirmed the diagnosis: spindle B cell melanoma iridis – pT1b/. No further treatment was indicated.

On the first postoperative examination the patient's visual acuity was l.0. Intraocular pressure (IOP) was 15 mm Hg. The patient was satisfied with cosmetic result and had no glare symptoms.

Discussion

The majority of iris melanomas are composed either exclusively of spindle melanoma cells, as it was in our patient, or of an admixture of spindle melanoma cells and benign nevus cells[3]. Most of the remaining iris melanomas consist of an admixture of spindle and epitheloid melanoma cells. Relatively few iris melanomas are composed exclusively of epitheloid melanoma cells.

The usual symptom of iris melanoma is a visible spot on the iris or discoloration of the iris in one eye. Many patients with iris melanoma have no symptoms, and the lesion in these patients usually is detected on routine eye examination. Less common symptoms are increased IOP, impaired vision, or spontaneous hyphema.

Our patient noticed the spot in his eye but had no symptoms either before or during the 3-year follow up.

Although its location makes the iris easily accessible for examination, differentiation of melanocytic malformations such as nevi or melanomas is difficult[4]. The decision to treat or observe a suspect iris melanoma is currently based on clinical features. Clinicopathologic studies have determined that many excised iris tumors are nevi and borderline lesions rather than frank melanoma. Identification of those clinical features suspect of melanoma remains challenging[3,5-8].

Documented growth of a suspect lesion represents an important diagnostic feature. However, previous studies have reported that even nevi may display slow growth that does not necessarily indicate malignancy[3,5,9]. Some studies of the pathogenesis of melanoma where slow growth phase of benign or precursor lesions occurs, being replaced by entry into an accelerated growth phase, report on an association with malignant progression[10,11].

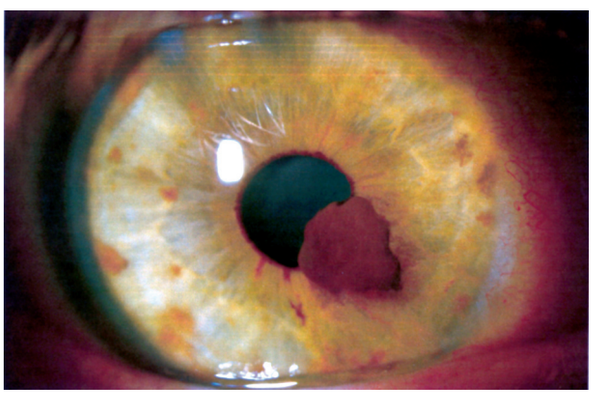

Fig. 1. Lesion three years after first examination: initial signs of progression.

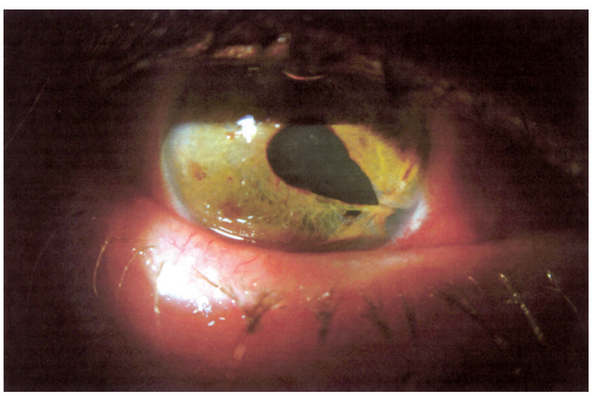

Fig. 2. One week postoperatively: good cosmetic result.

These findings highlight the need of regular lifelong review of these lesions with careful biometry and documentation of the relation of the tumor to the surrounding landmarks (especially the angle structures) to enable timely detection and management of melanomas displaying malignant change.

Iris melanomas display a variety of growth patterns. Two main categories include nodular or diffuse growth patterns. Iris melanoma usually grows locally into the anterior chamber or along the iris surface, and commonly invade the anterior chamber angle and anterior ciliary body by local extension. „Ring melanoma" is a term used to describe a tumor which grows around the iris in a circular shape. These tumors usually display a diffuse growth pattern[12].

In our patient, melanoma showed nodular growth pattern and was located near pupilar iris edge, and did not spread to the iridocorneal angle.

Most of the studies consider that iris melanomas should be diagnosed clinically in the presence of an iris melanocytic lesion that has locally replaced iris stroma, and/or is >3 mm in diameter or 1 mm thick and/or is associated with one or more of the following features including prominent vascularity, ectropion uveae, secondary cataract, secondary glaucoma, and evidence of documented growth[9,13,14].

Multiple studies have sought to clarify risk factors for malignancy, which include photographic documentation of growth, size (>3 mm diameter, >1 mm thick), glaucoma and/or pigment dispersion, invasion, prominent vascularity, hyphema and ring growth[12].

Our patient underwent operation after we had noticed initial signs of tumor growth (after three years): high pigmentation, 3x3 mm in size, slightly but evidently more prominent nodule, and presence of surface vascularity.

The requirement for the development of an intrinsic vasculature to supply an increasing tumor mass is well recognized. These vessels often have a disorganized structure and usually lack barrier function, which may be useful diagnostically in distinguishing these vessels from those seen in benign conditions[15-17]. A number of studies have reported that documented growth, basal diameter >3 mm, abnormal vasculature, pigment dispersion, satellite lesions, and tumor related symptoms are highly suspect of melanoma, whereas ectropion uveae, anterior chamber angle involvement, slow growth, and even glaucoma do not always clearly distinguish reliably between nevi and melanomas[9,14]. These features associated specifically with a more aggressive epitheloid or mixed cell histology remain to be determined.

Conway *et al.*[2] identified three features which were associated with the epitheloid component histologically: rapid growth, prominent tumor vessels and heterogeneous pigmentation. Taken in association with other features, these factors may have an important role in enhancing diagnostic accuracy for more aggressive iris melanomas[13].

Treatment options for iris melanoma include observation, excision, enucleation, and plaque radiotherapy. It usually consists of excision of the tumor, while enucleation today is occasional and required for large melanomas that cannot be resected. On resecting iris melanoma, careful assessment for iridocorneal angle involvement is important in treatment planning[2].

Over the past 20 years, advances in microsurgical technique, such as introduction of the partial lamellar flap, have improved access to the iris, anterior chamber angle, and ciliary body[18,19]. These techniques have allowed for more precise tumor resection to become possible, minimizing trauma to the eye and greatly reducing the complication rate.

Plaque radiotherapy is a newer treatment for iris melanoma. Because of anterior location of iris melanoma, radiation retinopathy is not common after treatment, but cataract and iritis are very common adverse reactions of radiotherapy. Local radiotherapy and charged particle irradiation have recently received attention for the conservative management of iris melanomas. The complications of ocular radiotherapy are well known and it is recognized that many of these have a considerably delayed onset, often years after the initial treatment. However, few studies have reported on the long term outcome of these therapies for iris melanoma[20-23].

Iris melanoma may present with elevated IOP, although the majority do not. Causes of increased IOP are still controversial, but include the following: angle invasion by tumor, pigment dispersion, anatomic angle closure by tumor (rare), and neovascularization following plaque radiotherapy.

Elevated IOP can be treated by topical or oral medications. Treatment of the iris melanoma may help reduce IOP as well. Performing argon laser trabeculoplasty (ALT) is controversial, since it may cause dispersion of tumor cells. Some authors have suggested performing ALT well away from the tumor. Cyclodestructive procedures are also controversial. Filtration surgeries are

contraindicated because they have been associated with metastasis[12].

Metastasis is unusual in iris melanoma. It is reported that, overall, 3%-5% of iris melanomas will metastasize after 10 years[12], whereas for mixed and epitheloid tumors, Geisse and Robertson have found that 11% and 7% respectively will eventually metastasize[5].

Recognition of clinical features associated with these more aggressive melanomas may have implications for earlier intervention.

Most recurrences occurred within the first 3 years after treatment, although some authors have reported on few patients that recurred up to 10 years later, indicating that prolonged postoperative follow up of these patients is necessary[24]. However, only a limited number of studies have reported on long term outcomes of such an approach for the management of iris melanoma[8,13,25-29].

Complications following iris melanoma resection including hemorrhage, vitreous loss, dislocated lens, cataract, iridocyclitis, macular edema, secondary glaucoma, and retinal detachment have been reported[8,25-29].

Recent studies report postoperative glare as the most common postoperative complication, which occurs in about a quarter of patients or just over, due to pupil reconstruction which has been performed in suitable cases in the past 10 years (usually less than 3-4 clock hours resected).

Conway *et al*. report that all patients undergoing local resection had postoperative vision no less than 0.1 and 78.6% had a visual acuity 0.5 or greater within the first postoperative year. The proportion declined to 28 (66.7%) at more than 1 year postoperatively, in all patients due to cataract progression, which could be treated surgically[2].

We performed iridectomy with sclerotomy and iris reconstruction, and had no intraoperative or postoperative complications. One week after surgery the patient's visual acuity was l.0. IOP was l5 mm Hg. The patient was satisfied with the cosmetic result and had no glare symptoms.

In conclusion, we can say that management decisions for iris melanoma depend on clinical features. As the lesion can persist without change for years and then turn into rapid growth phase, thorough follow up with photo documentation is necessary.

In the presence of two or more signs of malignancy, early intervention may be needed. When surgical treatment is undertaken, local resection achieves long term tumor clearance with an acceptable morbidity. Prolonged postoperative follow up is also necessary.

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Sažetak

MELANOM ŠARENICE: PRIKAZ SLUČAJA

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Premda je šarenica svojim smještajem lako dostupna pregledu, razlikovanje melanocitnih lezija kao što su nevus i melanom može biti teško. Neprepoznat melanom može dovesti do ozbiljnih okularnih i sustavnih komplikacija. Kod pravodobno postavljene dijagnoze prognoza je dobra. Odluka o daljnjim postupcima ovisi o kliničkoj slici. U većini slučajeva lokalna resekcija te pažljiv pristup iridokornealnom kutu, ako je zahvaćen, ne dovodi do recidiva bolesti te pruža zadovoljavajuće nizak pobol. Prikazujemo slučaj 52-godišnjeg muškarca koji je došao na pregled jer je primijetio pigmentiranu leziju na šarenici desnog oka.

Nakon temeljitog lokalnog i sustavnog pregleda, poslije tri godine praćenja učinjena je iridektomija sa sklerotomijom te rekonstrukcija šarenice kako bi se spriječila poslijeoperacijska fotofobija. Bolesnik je bio zadovoljan i funkcionalnim i kozmetskim rezultatom. Nije bilo indikacija za bilo kakav daljnji postupak liječenja.

Ključne riječi: *melanom šarenice*, *iridotomija*