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ПРИКАЗ БОЛЕСНИКА / CASE REPORT
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Iris melanocytoma

Anica Bobić Radovanović[1,2], Bojana Dačić Krnjaja[1,2], Vesna Jakšić[1,3]

[1]University of Belgrade, School of Medicine, Belgrade, Serbia;
[2]Clinic for Eye Diseases, Clinical Center of Serbia, Belgrade, Serbia;
[3]Clinic for Eye Diseases, Clinical Center Zvezdara, Belgrade, Serbia

SUMMARY

Introduction Iris melanocytoma (IM) is a rare benign tumor, but unavoidable in differential diagnosis of pigmented iris lesions. According to the best knowledge of the authors it is for the first time in Serbia that a well-documented case of IM is presented and that the problem of this tumor is discussed.

Case Outline In the left eye of a 47-year-old white female at the iris in a six o'clock position, a highly pigmented, dome shaped lesion with a crater-like cavity in the center and with feathery margins was noticed. There were no signs of infiltration of surrounding tissue or intrinsic vessels and the lens was clear. Visual acuity and intraocular pressure were normal. An ultrasound biomicroscopy (UBM) revealed a well-defined lesion with high internal reflectivity, with a base diameter of 1.25 mm and a thickness of 0.80 mm in the periphery, and 0.53 mm in the central part. The diagnosis of IM of the left eye was established and regular checkups were performed for ten years. No changes in clinical or UBM presentation were established.

Conclusion Awareness of clinical presentation of IM is most important for correct diagnosis. Ultrasound biomicroscopy is a useful diagnostic procedure in the following up of IM.

Keywords: iris melanocytoma; clinical presentation; ultrasound biomicroscopy

INTRODUCTION

For most ophthalmologists, melanocytoma is a rare tumor with distinguishing clinical characteristics located on or adjacent to the optic nerve head. However, this tumor can also be located in choroid, ciliary body and iris as well.

Iris melanocytoma (IM) is a benign tumor, a rare variant of iris nevus, with a distinctive clinical and histopathologic presentation.

IM is an exceedingly rare lesion. In a clinic-based series of 3,680 iris tumors collected over a 40-year period, only 68 lesions were diagnosed as melanocytoma [1]. Among 200 patients with suspected iris melanoma, only one had IM, and among approximately 1,400 patients with suspected iris nevus, only 3% were classified as having melanocytoma [2, 3]. In a series of 189 specimens of clinically suspected iris or ciliary body melanoma, 10 were melanocytoma [4].

IM can be seen in patients of all ages, but it is usually diagnosed in adults. Less than 10% of reported cases were in children younger than 10 years of age [3]. In a group of 17 patients with anterior uveal melanocytoma, median age on presentation was 55 years [5]. Approximately two thirds of reported cases of IM were females.

Clinical presentation of IM is distinctive. As a rule, a lesion is asymptomatic. A darkly pigmented, elevated, dome shaped lesion with feathery margins is usually present in the iris root in the inferior quadrant. A crater like cavity can be seen at the lesion's surface. Pigment dispersion can be present. There is no sign of infiltration of surrounding structures, no intrinsic vascularization, nor partial cataract.

Some changes can be observed during certain period of time as a part of natural evolution of a lesion.

IM demonstrates a tendency toward spontaneous necrosis. Cells from necrotic lesion and pigment containing macrophages can be seen on the iris surface and in the anterior chamber angle. New tumor seeds develop in more than 60% of IM at 10 years [3]. Pigment dispersion can cause secondary, melanocytomalytic, glaucoma. Increased intraocular pressure can be observed in 11% of patients with IM at 10 years [3].

IM can show progressive growth. Demirci et al. [3] estimate that tumor growth will be observed in nearly half of IM in 10 years. Malignant transformation of IM is extremely rare [6].

Histopathologic presentation of IM is characteristic. Plump, polyhedral cells are densely packed with melanin granules. After bleached preparation, relatively small nuclei with few nucleoli can be seen. Electron microscopy shows a large number of giant melanosomes in the cytoplasm [7].

CASE REPORT

A 47-year-old white female visited the ophthalmologist for a regular presbyopic correction in 2005. She did not have any ocular symptoms except reading problems. A slit lamp examination revealed a dark brown lesion located in a six o'clock position at the iris root of the left eye (Figure 1). The lesion was highly pigmented, dome shaped with a crater-like cavity in the center and with feathery margins. There were

Correspondence to:
Anica BOBIĆ RADOVANOVIĆ
Daničareva 63, 11000 Belgrade
Serbia
chjserbia@hotmail.com

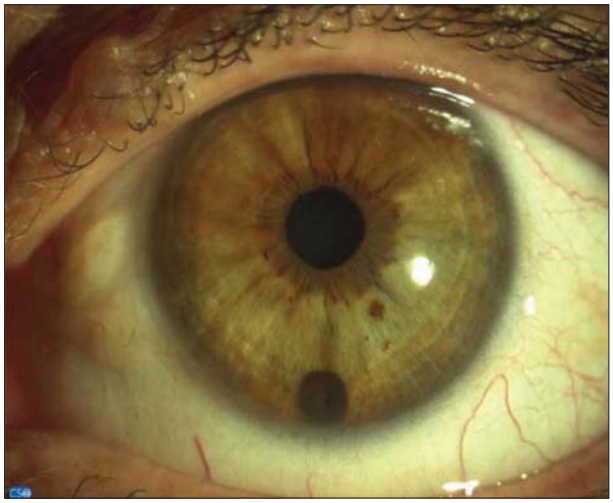

Figure 1. Clinical presentation of iris melanocytoma

Figure 2. Ultrasound biomicroscopy of iris melanocytoma

no signs of infiltration of surrounding tissue or intrinsic vessels. A gonioscopy showed an open angle of the anterior chamber, with a discrete dispersion of pigment in the lower parts. Transillumination demonstrated a pathologic shadow strictly on the iris. The lens was clear. Visual acuity was 20/20 in both eyes, and intraocular pressure was normal, 17 mmHg in both eyes. Complete examination showed no other functional or morphologic problems in either eye or orbit.

An ultrasound biomicroscopy (UBM) examination was performed. A nodular iris lesion with well-defined edges and a more or less regular surface with excavation in the middle zone was found (Figure 2). The diameter of the lesion was l.25 mm. The thickness was 0.80 mm in the periphery and 0.53 mm in the central part of the lesion. Internal tumor reflectivity was high. The angle of the anterior chamber was spared.

The diagnosis of iris melanocytoma of the left eye was established and regular checkups were recommended.

The patient was followed up with every six months for five years, and after that once per year. During a ten-year period no changes in clinical or UBM presentation were established.

DISCUSSION

According to our best knowledge, for a first time in our country, we present a well-documented case of IM and discussed the problem of this lesion.

The clinical presentation of this reported case was characteristic for IM: a middle-aged female with an asymptomatic, darkly pigmented dome-shaped lesion with central crater-like excavation in the inferior iris quadrant.

The majority of cases are diagnosed clinically based on the characteristic clinical presentation of IM. UBM and anterior segment optical coherence tomography (AS-OCT) are useful diagnostic procedures. The comparison between UBM and AS-OCT for imaging of 200 tumors of the anterior segment showed that UBM provided better imaging of pigmented iris tumors [8]. Typically, IM is a lesion of high internal reflectivity on a UBM image. This method is also important in following up IM as well. In select cases, there is a need for histopathological diagnosis. Fine needle biopsy is a useful but not a completely accurate procedure [9]. Excisional biopsy is both a diagnostic and therapeutic measure.

In our case, a diagnosis of IM was based on the clinical presentation and a UBM image of the lesion.

IM presents a problem in differential diagnosis of pigmented iris lesions, especially with iris melanoma. Documented tumor growth and secondary glaucoma can be seen in both lesions, but visible intrinsic vessels, corectopia, ectropion iridis and sector cataract suggests iris melanoma. Shields et al. [10] observed ectropion iridis in 44%, intrinsic vascularization in 43%, and sector cataracts in 14% of iris melanomas.

A cautious observation of a lesion is the treatment of choice for most IMs. In cases of unusual clinical presentation, documented rapid tumor growth or secondary glaucoma surgical treatment is indicated. Usually, local resection is sufficient, but in some cases enucleation is necessary.

In the presented case, a ten-year follow-up period confirmed that the diagnosis and the treatment of IM were correct. It is by all means necessary to continue with regular checkups.

IM is a rare benign tumor, but unavoidable in differential diagnosis of pigmented iris lesions. Awareness of clinical presentation of IM is the most important factor for correct diagnosis. Cautious observation of a lesion is the treatment of choice for most cases.

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Меланоцитом дужице

Аница Бобић Радовановић[1,2], Бојана Дачић Крњаја[1,2], Весна Јакшић[1,3]

[1]Универзитет у Београду, Медицински факултет, Београд, Србија;
[2]Клиника за очне болести, Клинички центар Србије, Београд, Србија;
[3]Клиника за очне болести, Клиничко-болнички центар „Звездара", Београд, Србија

КРАТАК САДРЖАЈ

Увод Меланоцитом дужице је редак, бениган тумор на који се мора мислити у оквиру диференцијалне дијагнозе пигментованих лезија дужице. Ово је, према доступним сазнањима аутора, први пут да се на нашим просторима приказује добро документовани случај меланоцитома дужице и да се дискутује о том проблему.

Приказ болесника На позицији 6 сати, на дужици левог ока 47-годишње жене беле расе установљена је изразито пигментована, ограничена лезија у облику куполе са кратерастим удубљењем на врху. Није било знакова инфилтрације околних структура, неоваскуларизације, ни замућења сочива. Видна оштрина и интраокуларни притисак имали су физиолошке вредности. Ултразвучном биомикроскопијом (УБМ) је утврђена јасно ограничена лезија високе рефлективности, пречника базе од 1,25 *mm*, дебљине од 0,8 *mm* на периферији и 0,53 *mm* у централном делу. Постављена је дијагноза меланоцитома дужице левог ока, а редовни контролни прегледи вршени су током десет година. Нису забележене промене ни у клиничкој, ни у УБМ презентацији лезије.

Закључак Добро познавање клиничке слике меланоцитома дужице од кључног је значаја за постављање исправне дијагнозе. УБМ је корисна дијагностичка метода. Меланоцитом ириса је бенигни тумор који обично не захтева никакву терапију, али је редовно и пажљиво клиничко праћење лезије обавезно.

Кључне речи: меланоцитом дужице; клиничка слика; ултразвучна биомикроскопија

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