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CHOROID MELANOMA, DEVELOPED FROM NEVUS

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- \Rightarrow It is known that 25% of choroidal nevi are suspicious, and the risk of their malignization is 2–13% with a tendency to increase with longer follow-up. It is assumed that 5.8% progressing choroidal nevi malignize within 5 years, and 13.9% of within 10 years. In the article, a clinical case of choroidal melanoma development 5.5 years after the detection of progressing nevus in a patient who refused to be followed-up is described. In the present case, there was a combination of two risk factors of nevus malignization, which significantly increases the likelihood of such an outcome. Long-term follow-up of patients is required from the first day of diagnosis "choroidal nevus with signs of progression".
- *★ Keywords:* choroidal melanoma; progressing choroidal nevus; choroidal nevus malignization; optical coherent tomography.

МЕЛАНОМА ХОРИОИДЕИ, РАЗВИВШАЯСЯ ИЗ НЕВУСА

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- \Leftrightarrow Известно, что подозрительными являются 25 % невусов хориоидеи, а риск озлокачествления их составляет 2-13 % с тенденцией к увеличению по мере удлинения сроков наблюдения. Считают, что в течение 5 лет переходу в злокачественную опухоль подвергается 5,8 %, а в течение 10 лет 13,9 % прогрессирующих невусов хориоидеи. В статье описан клинический случай развития меланомы хориоидеи, через 5,5 лет после выявления, прогрессирующего невуса у пациента, отказавшегося от наблюдения. В представленном случае имело место сочетание двух факторов риска озлокачествления невуса, что существенно повышает вероятность такого исхода. Необходимо длительное диспансерное наблюдение за пациентами с первого дня постановки диагноза «невус хориоидеи с признаками прогресии».
- *★ Ключевые слова*: меланома хориоидеи; прогрессирующий невус хориоидеи; озлокачествление невуса хориоидеи; оптическая когерентная томография.

INTRODUCTION

Differential diagnosis of progressive choroidal nevus (PCN) and initial choroidal melanomas (CMs) represents an important and complex issue of modern ophthalmic oncology. About 25% of PCNs are known to be considered suspicious [1], and the risk of malignancy is 2% to 13% with a tendency to increase as the follow-up period increases [1–4]. According to the literature, 5.8% of PCNs become malignant within 5 years, and 13.9% become malignant within

10 years [5]. The most significant clinical risk factors for nevus transformation into CMs include visual impairment, emergence of an orange pigment on the nevus surface, increase in its thickness and diameter during follow-up, appearance of a "void" symptom based on ultrasound (US) examination, and presence of subretinal fluid based on optical coherence tomography (OCT) [2, 5]. In the absence of risk factors for 5 years, malignancy in 1.1%—3% of cases is possible [5—8]. The risk of PCN conversion

to the malignant stage can reach 55% when more than 2 factors are combined. In this regard, according to C.L. Shields et al. [5, 6], such PCNs can be regarded as initial melanomas.

Herein, our own clinical case is illustrated.

Patient K., 58 years old, presented to the Research Institute of Eye Diseases with complaints of floating opacities in both eyes. At the initial examination, visual acuity (VA) of both eyes was 0.9 without correction, with sph-0.5 VA was 1.0. Intraocular pressure was 18 mm Hg in both eyes, and perimetry revealed no pathological changes.

In the anterior segment of both eyes, there were no pathological changes; there was destruction of the vitreous body.

When examining the fundus of the right eye, a pigmented lesion with clear boundaries and drusen on the surface was revealed in the temporal paramacular area (Figure 1, *a*). The left eye fundus had no local pathological changes.

Ultrasound (US) examination of the right eye revealed an additional shadow with a prominence of 2.32 mm and a diameter of 6.36 mm.

The patient underwent fluorescein angiography to clarify the diagnosis. In the area of the pigmented lesion, blocking of background choroidal fluorescence was visualized in all phases of the examination. Spotted hyperfluorescence is visible, which intensity increased during the examination, in the center of the focus starting from the arteriovenous phase. In the late phase (40 minutes after the administration of the dye), confluent hyperfluorescence with point regions of brighter "hot spot" hyperfluorescence persisted in this zone (Figure 1, b-d).

The lesion identified was regarded as a progressive nevus, and follow-up was recommended for the patient.

The examination after 3 months revealed no dynamics in the focus status; therefore, repeated examination after 6 months was recommended. However, the patient refused to be monitored and contacted the Research Institute of Eye Diseases only after 5.5 years, when visual impairment of the right eye occurred. At the examination, VA of the right eye was 0.7 incorrigible, VA of the left eye was 1.0, and intraocular pressure was 18 mm Hg in both eyes. Perimetry revealed paracentral absolute scotoma in the visual field of the right eye. Anterior segment of both eyes had no pathological changes, and there was destruction of the vitreous body.

At ophthalmoscopic examination of the right eye, in the temporal paramacular area, there was a protrud-

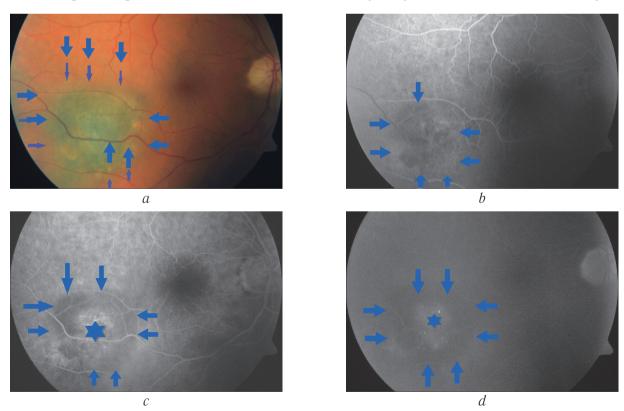


Fig. 1. Fundus photo (a), early (b), middle (c) and late (d) stages of fluorescein angiography at the primary patient examination. Arrows indicate the nevus borders, \star – hyperfluorescent nevus center

Рис. 1. Фото глазного дна (a), ранняя (b), средняя (c) и поздняя (d) фазы флюоресцентной ангиографии при первичном обращении пациента. Границы невуса указаны *стрелками*. ★ — зона гиперфлюоресценции в центре невуса

ing, unevenly pigmented focus of irregular shape with fuzzy boundaries, reaching the fovea, with an orange pigment field at the edges of the lesion (Figure 2). The fundus of the left eye had no local pathological changes.

Ultrasound examination of the right eye revealed an additional shadow with prominence of 3.36 mm and a diameter of 10.1 mm. Thus, over 5.5 years, the diameter of the tumor increased by 58.81% and its prominence increased by 44.83%, which is not typical for progressive nevus.

To clarify the diagnosis and determine the management approach, OCT was performed, including in the enhanced depth imaging mode. Two-apices elevation of the choroidal complex with an intense hyperreflective structure under the Bruch's membrane was revealed, which blocked the signal from the underlying tissues. In the central and most protruding part of the focus, areas of Bruch's membrane integrity damage were revealed along with an extended slit-like detachment of the pigment epithelium with moderately hyperreflective content above it, with single cavities and point intensely hyperreflective inclusions. The differentiation of the overlying retinal layers was impaired. There was a detachment of neuroepithelium with signs of edema and rupture of photoreceptors between the apices of the lesion. Along the lower edge of the focus, there were single cavities directly under the Bruch's membrane (Figure 3). The signs revealed by OCT correspond to the presentation of choroidal melanoma [9, 10].

Comparison of the fluorescein angiography results performed at the initial visit with the results of OCT performed after 5.5 years indicated that the confluent hyperfluorescent zone corresponded to the zone of the long slit-like detachment of the pigment epithelium with a moderately hyperreflective content on the surface, which may retrospectively indicate the presence of detachment of the retinal pigment epithelium at the time of fluorescein angiography. There were zones of retinal pigment epithelium defects in places of bright "hot spot" hyperfluorescence.

Impairment of visual functions, increase in the size of the lesion, and OCT data enabled to establish the diagnosis of choroidal melanoma that developed from PCN.

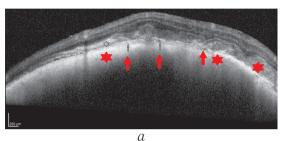
After a general examination confirming the location of the tumor, the patient underwent brachytherapy (apical dosage of 127 Gy). A positive dynamic was noted 4 months after brachytherapy, as the prominence and diameter of the CM decreased to 1.08 mm and 7.93 mm, respectively.

About 20 months after brachytherapy, OCT revealed a scar with pigment hyperplasia in the central zone of the fundus in the irradiation zone (Figure 4),



Fig. 2. Fundus photo after 5.5 years after first patient's visit

Рис. 2. Фото глазного дна пациента через 5,5 лет после первичного обращения



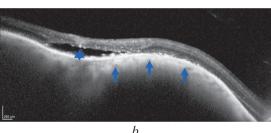


Fig. 3. OCT horizontal (a) and vertical (b) scans across the tumor: $a - red \ arrows -$ slit-like pigment epithelium detachment, ★ - hyperreflective content above the pigment epithelium detachment; $b - blue \ arrows -$ single cavities under the Bruch's membrane, ★ - neuroepithelium detachment between the tumor apices

Рис. 3. Оптическая когерентная томография: горизонтальный (а) и вертикальный (б) сканы через образование: а — красные стрелки — щелевидная отслойка пигментного эпителия, ★ — гиперрефлективное содержимое над отслойкой пигментного эпителия; b — синие стрелки — единичные полости под мембраной Бруха, ★ — отслойка нейроэпителия между горбами опухоли

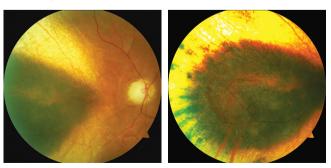


Fig. 4. Fundus photo, 20 months after brachytherapy

Рис. 4. Фото глазного дна через 20 мес. после брахитерапии

with prominence of 0.47 mm according to US. Thus, complete resorption of melanoma was recorded after brachytherapy. According to MRI of the abdominal organs and CT of the chest organs, no pathology was detected.

Visual acuity of the right eye was 0.01 (incorrigible). Perimetry revealed narrowing of the visual field from the nasal side with the fixation point involvement.

Thus, the presented clinical case confirmed the possibility of CM development from a progressing nevus and the opinion established in the literature that the combination of 2 or more risk factors (in our case, an increase in the size of the focus and the emergence of an orange pigment), detected during the follow-up, indicates the conversion of a nevus into a melanoma. The conversion of PCN in this case that presented as the initial melanoma after 5.5 years necessitates long-term follow-up of patients from the day the diagnosis of choroidal nevus with signs of progression was established.

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