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Scientific article



Differential diagnosis of peripheral exudative hemorrhagic chorioretinopathy and neoplasm of the choroid (clinical case)

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Peripheral exudative hemorrhagic chorioretinopathy is a relatively rare and difficult to diagnose disease. This condition is clinically similar to choroidal melanoma, which is why it is called “pseudomelanoma”. An erroneous diagnosis of choroidal melanoma can lead to the wrong choice of aggressive treatment tactics. The aim of this work was to present a case of differential diagnosis of suspected neoplasm of the choroid with peripheral exudative hemorrhagic chorioretinopathy. The described clinical case demonstrates characteristic clinical picture and results of ultrasound with Doppler mapping, spectral optical coherence tomography, optical coherence tomography–angiography, scanning laser ophthalmoscopy for this condition, as well as important differential diagnostic signs of choroidal melanoma. Complaints, history, clinical picture and the results of instrumental examinations were characteristic of peripheral exudative hemorrhagic chorioretinopathy and allowed to exclude the diagnosis of choroidal neoplasm. Pathogenetic treatment (intravitreal injection of anti-VEGF agents) and observation were recommended to the patient, since this disease often affects both eyes. The main differential diagnostic criterion for suspected choroidal melanoma is Doppler ultrasound imaging. In difficult clinical cases, structural optical coherence tomography, optical coherence tomography–angiography, and scanning laser ophthalmoscopy provide valuable additional information for verifying the diagnosis.

Keywords: peripheral exudative hemorrhagic chorioretinopathy; polypoid choroidal vasculopathy; choroidal melanoma; ultrasound mapping; scanning laser ophthalmoscopy; optical coherence tomography.

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Научная статья

Дифференциальный диагноз периферической экссудативной геморрагической хориоретинопатии и новообразования сосудистой оболочки (клинический случай)

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Периферическая экссудативная геморрагическая хориоретинопатия — относительно редкое и трудно диагностируемое заболевание. Данная патология по клинической картине схожа с меланомой хориоидеи, из-за чего её называют «псевдомеланомой». Ошибочно установленный диагноз меланомы хориоидеи может привести к неправильному выбору агрессивной тактики лечения. Цель настоящей работы — представить случай дифференциальной диагностики подозрения на новообразование сосудистой оболочки с периферической экссудативной геморрагической хориоретинопатией. Описанный клинический случай демонстрирует характерные клиническую картину и результаты ультразвукового исследования с доплеровским картированием, спектральной оптической когерентной томографии, оптической когерентной томографии-ангиографии, сканирующей лазерной офтальмоскопии для данной патологии, а также важные дифференциально-диагностические признаки меланомы хориоидеи. Данные жалоб, анамнеза, клинической картины и результаты инструментальных исследований были характерны для периферической экссудативной геморрагической хориоретинопатии и позволили нам исключить диагноз новообразования сосудистой оболочки. Пациентке было рекомендовано патогенетическое лечение (интравитреальное введение ингибиторов ангиогенеза) и наблюдение, так как нередко данное заболевание поражает оба глаза. Основным дифференциально-диагностическим критерием при подозрении на меланому хориоидеи является ультразвуковое исследование в режиме доплеровского картирования. В трудных клинических случаях структурная оптическая когерентная томография, оптическая когерентная томография-ангиография и сканирующая лазерная офтальмоскопия дают ценную дополнительную информацию для верификации диагноза.

Ключевые слова: периферическая экссудативная геморрагическая хориоретинопатия; полипоидная хориоидальная васкулопатия; меланома хориоидеи; ультразвуковое доплеровское картирование; сканирующая лазерная офтальмоскопия; оптическая когерентная томография.

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BACKGROUND

Peripheral exudative hemorrhagic chorioretinopathy (PEHC) is a rare degenerative exudative hemorrhagic disease of the peripheral retina occurring in patients aged >65 years (70% of the cases report female patients) [1–5]. Some authors name this disease “pseudomelanoma,” as the differential diagnostics between a choroidal melanoma and PEHC often causes difficulties in the practice of ophthalmology [3]. PEHC may be clinically accompanied by pathological changes along the retinal vessels in the form of subretinal hemorrhages or exudation, the presence of solid exudates, retinal pigment epithelium detachment, and in rare cases, hemophthalmos may develop. More often, changes are detected in the retinal periphery, in the lower temporal quadrant [2, 3]. In the early stages, the disease is asymptomatic, the visual acuity remains quite high, but with the progression of the pathological process in the form of hemorrhages, spread of exudation and pigment epithelium detachment toward the macula, visual acuity decreases significantly [2–5]. Due to the great similarity of the ophthalmoscopic presentation, this condition must be differentiated from a choroidal melanoma. An erroneous diagnosis may lead to the incorrect choice of the aggressive treatment approach (thermotherapy, radiation methods, and often, enucleation). Hence, additional research methods, such as the Doppler ultrasound, optical coherence tomography (OCT), OCT angiography, and scanning laser ophthalmoscopy, may be important for the differential diagnosis of the PEHC and the choroidal melanoma [1–7].

CLINICAL CASE

As a clinical example, the results of a comprehensive ophthalmological examination of a 66-year-old female patient V. are presented, who first came to the V.V. Volkov Ophthalmology Clinic of the Military Medical Academy with a previously established diagnosis of neoplasm of the left

eye choroid. Ultrasound examination of the eyeball was performed in a gray scale scanning (B-mode) and color Doppler mode, using the universal ultrasound system LOGIQ e (GE, China). Consequently, the absence or the presence of a vascular signal in the projection of the lesion was assessed, and its size and the localization were determined. A spectral OCT was performed on an RTVue-XR optical coherence tomograph (Optovue, USA), using a Cross Line structural scan (two 10 mm orthogonal scans) and an Angio Retina of 6-mm OCT angiography scan. The fundus photography and scanning laser ophthalmoscopy were performed using an AFC-330 fundus camera (Nidek, Japan) and F-10 confocal scanning laser ophthalmoscope (Nidek), respectively.

Upon admission, the patient complained of low visual acuity of the left eye. A decrease in visual acuity was noted over the past year. The best corrected visual acuity was 0.3. The patient reported hypertension as a concomitant systemic disease.

Indirect ophthalmoscopy along the course of the inferior vascular arcade visualized a mass oval-shaped lesion, dark, protruding into the vitreous, and 5×6 optic nerve head diameters in size. Accumulation of the subretinal material (organized blood) and blood was determined along the lower peripheral edge of the neoplasm, and hard exudates were noted along the central edge with the involvement of the macular center (Fig. 1).

An ultrasound examination with a Doppler mapping in the lower part of the eyeball detected a rounded lesion of a heterogeneous echo density, which was 0.32×0.80 cm in the size and had no vascular signal in the center of the lesion (Fig. 2).

The structural OCT determined a dome-shaped detachment of the retinal pigment epithelium blocking the passage of the OCT signal, accumulation of intraretinal fluid, and intraretinal hyperreflective inclusions, flat fibrovascular detachment of the retinal pigment epithelium with a “double layer” sign (Fig. 3, a). OCT angiography in the external retinal layers and choriocapillaris visualized

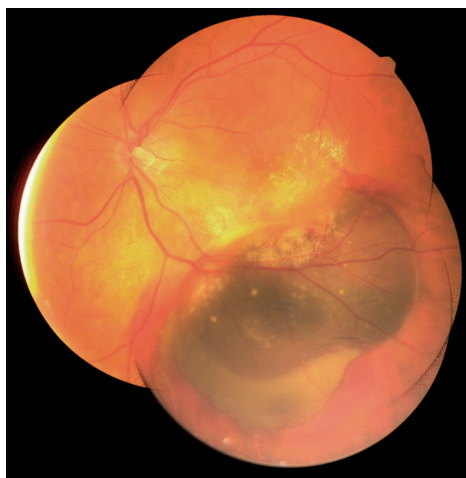


Fig. 1. Color fundus photography with oval mass

Рис. 1. Фотография глазного дна с объёмным образованием овальной формы

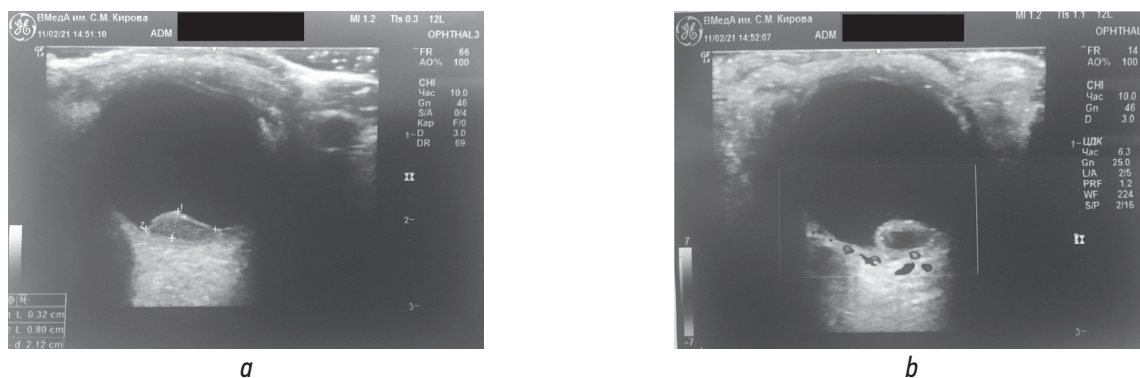


Fig. 2. Ultrasound procedure: *a* – B-scan mode; *b* – Doppler mapping mode

Рис. 2. Ультразвуковое исследование: *a* — режим В-сканирования; *b* — режим доплеровского картирования

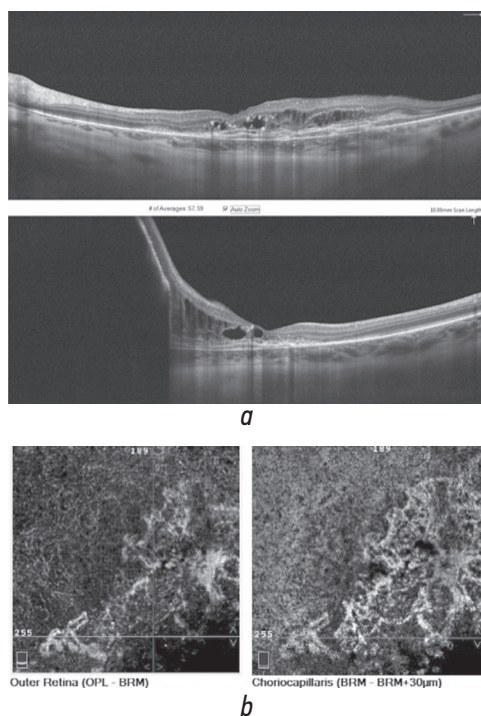


Fig. 3. Optical coherence tomography angiography: *a* – structural, retinal detachment is visualized; *b* – angiography, vascular network

Рис. 3. Оптическая когерентная томография-ангиография: *a* — структурная, визуализируется отслойка сетчатки; *b* — ангиография, сосудистая сеть

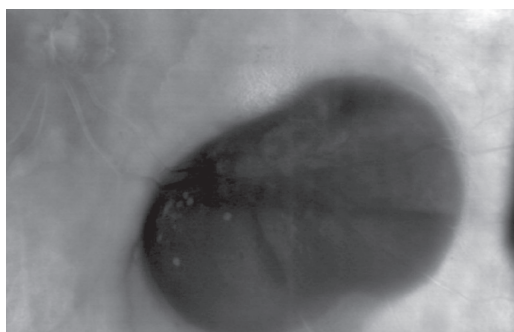


Fig. 4. Image of infrared scanning laser ophthalmoscopy in retro mode shows a round mass

Рис. 4. Инфракрасная сканирующая лазерная офтальмоскопия в ретро-режиме. Изображение округлого объёмного образования

the branching vascular network in the projection of the macular area (Fig. 3, *b*) [7].

The infrared scanning laser ophthalmoscopy in the retromode determined a rounded space occupying lesion completely blocking the signal from the underlying sclera. In contrast to the neoplasms of the choroid, this zone had the clear boundaries characteristic of hemorrhagic detachment of the retinal pigment epithelium (Fig. 4).

Based on a combination of multimodal imaging findings, PEHC was diagnosed in the patient. Due to the presence of an extensive subretinal hemorrhage as well as of a hemorrhage under the retinal pigment epithelium and the inappropriateness of the photodynamic therapy, intravitreal antiangiogenic therapy and routine case follow-up was recommended to the patient.

A choroidal mass is always a disturbing finding, since there are only a few conditions included into the range of the differential diagnosis of malignant neoplasms of the choroid. In addition to a melanoma and a choroidal metastasis, the mass lesion may be a giant nevus, choroidal hemangioma, vorticos vein, and the PEHC. Distinctive aspects of vorticos veins are relatively small size, and collapse at a pressure exerted on the eyeball (sclerocompression). The choroidal hemangioma has an almost exclusively central localization and a distinctive ophthalmoscopic presentation (round shape, red color, and smooth surface). Additionally, a specific presentation by the indocyanine green angiography is noted [8]. A giant nevus does not have exudative changes in the retina and is usually associated with the secondary changes in the retinal pigment epithelium, namely atrophy and drusen. Thus, in terms of the localization, volume, and presence of the exudative changes, PEHC could resemble melanoma or choroidal metastasis and pose a significant differential diagnosis problem. The low awareness of specialists about this condition causes an additional difficulty.

Although the exact genesis of the PEHC remains unknown, it is assumed that this condition is like a polypoid choroidal vasculopathy but differs by a peripheral localization. In our case, presence of the subretinal fresh and old blood, hard exudates, presence of intraretinal fluid in

the absence of a fluid under the retina, and presence of a branching neovascular network in the macula supported the diagnosis of the PEHC.

OCT revealed a high elevation of the retinal pigment epithelium, and although its content could not be confidently differentiated, in combination with the presence of a subretinal blood, its hemorrhagic nature could be assumed. This was also confirmed by the ultrasound data which demonstrated a hypointense signal in the neoplasm center.

The most important finding in this case was the neovascular network detected by the OCT angiography, which could not be due to a neoplasm and explained the presence of the intraretinal fluid and solid exudates.

Although one of the most informative methods for the differential diagnosis of choroidal melanoma and PEHC is undoubtedly indocyanine green angiography, it is not always available in the real clinical practice.

Choroidal melanoma could mimic PEHC, but the proposed examination methods will enable to establish with high probability the only correct diagnosis and prescribe pathogenetic treatment.

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CONCLUSION

This clinical case demonstrates the importance of considering PEHC as one of the points of the differential diagnosis for a suspected choroidal melanoma. A combination of the multimodal imaging methods detecting evidences of an active exudative process, associated with a choroidal neovascularization, may be useful in the relatively central location of the lesion.

ADDITIONAL INFORMATION

Author contributions. All the authors confirm that their authorship complies with the international ICMJE criteria, i. e., all the authors have made a significant contribution to the development of the concept, research, and preparation of the article, and have read and approved the final draft before its publication.

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