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Case Report

Choroid hemangioma: primary tumor or reactive angiomatosis?

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Abstract

Vascular tumors of the peripheral choroid are uncommon and may provide the clinician with a difficult diagnostic problem. In this article, we report 12 cases of eyes hemangiomas and 10 metastatic choroidal tumors to emphasize the significance of the initiating rol e of metaplastic and proliferative changes in the tapetoretinal zone that precede angiomatosis and cause it.

Keywords: angiomatosis, choroid hemangioma, metastatic choroidal tumors, morphology

Introduction

In clinical practice, choroid hemangioma is rare. Commonly, the disease is detected due to decreased vision as a result of changes around the tumor or as an accidental finding during a routine examination. The pathological process is almost always localized in one eye, most often on the left and on the temporal side, preferring the juxtapapillary zone, usually represented by a single node with fairly clear boundaries, round or oval, with a whitish-yellow or yellowish-pink.

Slate color or pronounced pigmentation around the tumor is due to the proliferation of retinal pigment epithelium over the newly formed vessels. The usual size of the tumor: diameter from 3 to 15 mm, the prominence from 1 to 5-6 mm. In our practice, there was a hemangioma in the peripheral zone with a stand up to 16 mm, with intense pigmentation, which ophthalmoscopically and according to the ultrasound study coincided with the picture of melanoma.

Secondary retinal detachment with an exudative component, characteristic of most hemangiomas, can significantly complicate the differential diagnosis of a tumor with melanoma or metastasis, which more often leads the doctor to undergo radical treatment, especially when involving the optic nerve, blindness, and fairly rapid progression of the neoplasm. In the later stages, neovascular glaucoma can develop, especially with total retinal detachment, which is also a reason for enucleation.

Diffuse tumors with different variants of Sturge-Weber syndrome, having diffuse red-orange thickening over a large extent in the post-equatorial zone, are less common. For differential diagnosis, ultrasound scanning, optical coherence tomography (OCT), angiography

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using fluorescein or indocyanine green, ophthalmoscopy in infrared light, MRI are used for differential diagnosis. Despite some obvious features characteristic of uveal hemangioma, its diagnosis is often difficult and requires differentiation, especially with non-pigmented or slightly pigmented melanoma [1-4].

From the position of pathomorphological diagnosis, the choroid hemangioma has never been of interest because of the obvious morphological picture, therefore, even in monographs and guidelines for the diagnosis of tumors, very little space is given to it. However, we want to draw attention to some circumstances that give reason to think about the nature of this angiomatosis.

First of all, it should be noted that this pathology appears as a tumor only within the framework of macroscopic observation, being a form familiar to many, including eye tumors. However, a histological examination in the choroid reveals only a complex of hypertrophy capillary vessels involved in general microcirculation. Moreover, there is no cell proliferation, which is an integral component of any tumor process, including such benign vascular tumors as capillary or cavernous hemangioma of other localizations.

It should be noted that similar extensive foci of choroidal angiomatosis occur with melanoblastomas and retinoblastomas both inside the tumor parenchyma and at the border with the tumor node. This is due to the fact that neoplastic growth initiates neovascularization , sometimes surpassing the tumor parenchyma in volume. In addition, the formation of new vessels in the choroid can occur not only in the form of a reaction to tumor growth but also during degenerative processes in the tapetoretinal zone as in age-related macular degeneration (AMD).

In all the situations noted, the formation of a hypertrophic vascular network in the choroid is secondary and is due to pathological processes both in the choroid and in the adjacent tapetoretinal zone. It is interesting that almost always with hemangiomas of the choroid, even when the distance of the node does not exceed 1-2 mm, one can observe retinal detachment and pronounced degenerative changes in the neuroepithelium, leading to visual impairment at such an early stage. On this basis, hemangiomas are superior to metastases, whose growth in the choroid, despite the malignant nature, is not accompanied by subretinal hemorrhages.

Moreover, uveal metastases are not characterized by such an extensive detachment and early degenerative changes in the retina that invariably accompany hemangioma and dissonant with the idea of it as a benign tumor. There is natural bewilderment in connection with the fact that a benign tumor has a more destructive effect on the structures of the tapetoretinal zone than metastases of malignant neoplasms. And this despite the fact that the hemangioma of the choroid is a complex of dilated vessels, which are included in the blood supply.

The foci of the proliferation of retinal pigment epithelium also deserve attention, which can often be observed over the hemangioma node. Only some morphologists drew attention to this pattern, although they did not attach much importance to it, considering it to be a concomitant reaction due to the growth of the tumor node in the choroid [5]. Perhaps that is why in later descriptions of the hemangiomas of the choroid, these changes are not discussed at all.

Case

A 46-year-old patient turned for the first time. On the left eye, massive retinal detachment, secondary glaucoma, lens opacification. O n OCT and ultrasound, an intraocular tumor was not detected. Given the general ophthalmic picture, it is recommended that enucleation be performed. A macroscopic examination of the distant eye in the peripapillary zone revealed a flattened tumor-like node with a distance of less than 1 mm and a length of up to 5 mm, the retina was totally detached, the exudate was yellowish-gray in color, very dense (easily peeled off).

Histological diagnosis: juxtapapillary hemangioma of the choroid, melanosis of the optic disc. In the tapetoretinal zone, fragments of fibrous-hyalinized tissue intimately connected with the inner surface of the choroidal node were revealed, the pigment epithelium at the top of the node is absent, and there are a few small proliferates from the pigment epithelium cells on the periphery. This case can serve as a demonstration of how pronounced pathological changes can be caused by the micronode of choroidal hemangioma. Similar examples cannot be found with metastases in the choroid, which confirms the idea expressed here: this pathological process does not begin with the formation of the angiomatous node in the choroid, but with those changes in the tapetoretinal zone that precede angiomatosis and cause it. For the same reason, choroidal angiomatosis can't explain the severity of pathological changes that do not correspo nd to the clinical picture of a benign tumor.

In order to analyze changes in the tapetoretinal zone, a histomorphological study of enucleated eyes with hemangioma and uveal metastases was performed. The histologic preparations of 12 hemangiomas and 10 metastatic choroidal tumors were studied. Both archival and current histomorphological material were used. For a more detailed study of the state of the tapetoretinal zone, serial sections were studied (in some cases, up to 35 sections from one node). Sections were made from paraffin blocks; standard hematoxylin-eosin staining was used. Histological examination data were compared with the clinical picture. In all cases, with hemangiomas of the choroid, changes in the pigment epithelium and a violation of the integrity of the Bruch's membrane were revealed, especially expressed in the

areas of the greatest distance of the choroid node. In some cases (6 observations), the tumor-like proliferation of pigment epithelium occurred over the top of the tumor node, sometimes with the formation of micronodules up to 500 μm in the largest size, not accessible for macroscopic imaging, but clearly visible microscopically [Figure 1].

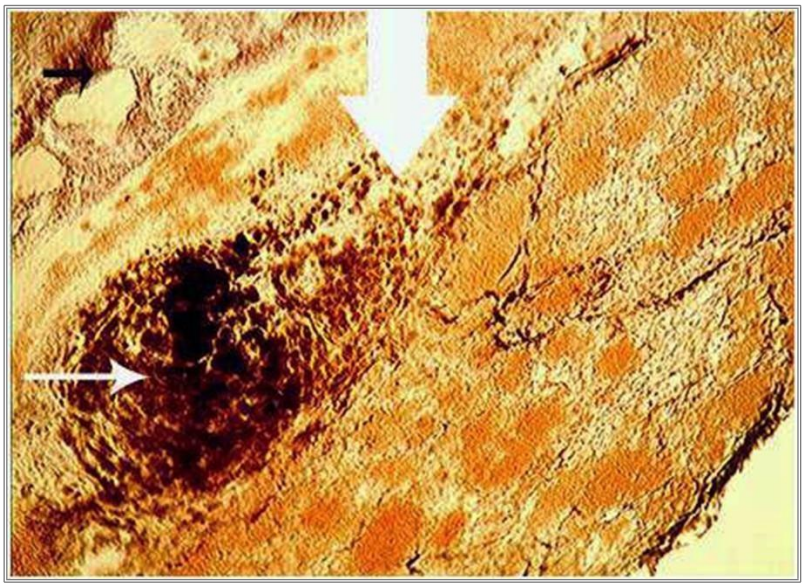

Figure 1. To the left of the complex of choroidal vessels is a visible pigment node (horizontal arrow). Bruch's membrane under it is not traced, and subretinal hemorrhage (vertical arrow) indirectly indicates a violation of its integrity. In the upper left corner (thin black arrow) there is a fragment of a detached retina with cystic degeneration. (Hematoxylin-eosin, x200).

Microscopic examination of the tapetoretinal zone with uveal metastases shows that retinal changes are often reduced to cystic degeneration of the inner layers and loss of cells in the pigment epithelium, and the patterns of proliferation and signs of metaplasia are not characteristic. It is important to note the mismatch between the degree of node extention and the severity of reti nal detachment and changes in the tapetoretinal zone, which occurs with hemangiomas. So, in none of the cases studied by us did the extinction of the angiomatous node exceed 4 mm (according to ultrasound and measurement on a micropreparation). Moreover, in 4 cases, the prominence was in the range of 1-2 mm. On the contrary, uveal metastases with an extension of less than 4 mm were not observed by us at all, which can be explained by their clinical indifference at this stage of growth. In this regard, one case we have observed deserves attention.

Discussion

It should be noted that in the cases noted, the proliferation foci represented by pigment epithelial cells exceeded the size of similar proliferates, which Reese and Jones proposed to consider as initial retinal pigment epithelium tumors [6,7]. The study of these authors can be considered a classic since that time tumor-like proliferations of the pigment epithelium without communication with intraocular melanomas or other pathological conditions in the eye have not been studied in such detail. In contrast to the aforementioned study, in all our cases, proliferative changes were accompanied by a violation of the integrity of the Bruch membrane. It is difficult to imagine such changes in the tapetoretinal zone above the tumor node as a secondary reaction. Indeed, an angiomatous node is only a hypertrophic vascular network, the elements of which can often be found on the periphery or in the center of the melanoma node growing in the choroid [Figure 2].

Such vascular patterns as in Figure 2 can be observed in cases of retinoblastoma implanted on Bruch's membrane. In such cases, vascular neoplasm can be considered as a reaction to invasion. From these positions, we propose to compare the patterns of the prolife ration of pigment epithelium in the tapetoretinal zone and the formation of blood vessels in the choroid in hemangioma. We have no reason to associate these patterns with any kind of hemangioma effect because there are no such changes with metastases in the choroid.

In those cases where tumor-like patterns could not be detected, however, other signs of metaplastic transformation of the retinal pigment epithelium were present. So, in one case, small foci of osteogenesis in the tapetoretinal zone were revealed against the background of the weakly expressed proliferation of pigment epithelium. Bruch's membrane was not detected in this area and osteogenic patterns could

be seen both intimately associated with the stroma of the choroidal hemangioma and among the chorioretinal fibrous tissue, in which altered pigment epithelial cells were present.

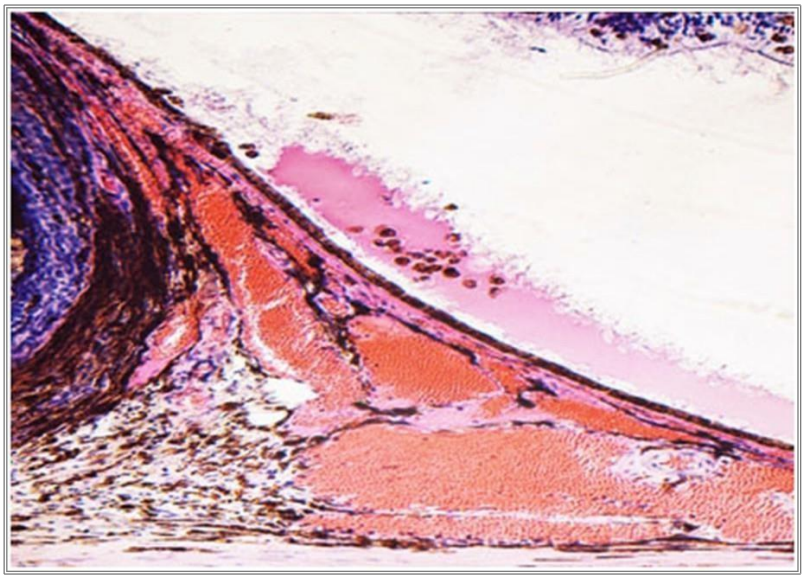

Figure 2. Hypertrophic vasculature in the area of the choroid adjacent to the melanoma node (left). The complex of dilated vessels resembles a hemangioma by its morphological structure. (Hematoxylin-eosin, x100).

Similar osteoplastic metaplasia of pigment epithelium has been well studied in post-traumatic processes of the eye, however, foci of osteogenesis in the tapetoretinal zone are not mentioned in the case of choroid hemangioma [1,8]. There is reason to believe that such metaplasia is the primary process associated with the proliferation of pigment epithelium, and it is the degenerative metaplastic changes in the pigment epithelium that lead to a violation of the integrity of the Bruch membrane, initiating angiomatosis in the choroid, and not vice versa. By the way, the sequence of changes - from degeneration in the tapetoretinal zone to neovascularization of the choroid is characteristic of the pathogenesis of AMD.

The view of hemangioma as an independent benign tumor formed in the choroid is dissonant with the clinical picture, in which exudative hemorrhagic retinal detachment is already present at an early stage, which can't happen without the corresponding destructive changes in the tapetoretinal zone. Such a violation requires the invasive properties of the tumor, which is not characteristic of benign hemangioma. Moreover, metastases in the choroid, the invasive properties of which are not in doubt, even with large sizes of tumor nodes, behave less aggressively against the tapetoretinal zone. All this allows us to express the point of view according to which the destruction of the Bruch membrane and the associated exudative hemorrhagic retinal detachment are not due to angiomatosis in the choroid, but are preceded by proliferative and metaplastic changes in the pigment epithelium, including its tumor-like proliferation.

This may be a developing neoplasm, which at an early stage of its development, without having its own stromal-vascular territory for the formation of a tumor node, can nevertheless lead to the destruction of Bruch's membrane and initiate angiogenesis in the choroid. The likelihood of such a relationship between the tumor and the choroidal vasculature is demonstrated by melanomas and retinoblastomas. One way or another, there is reason to continue research, including immunohistochemical one. The latter can help to identify neoplastic growth markers in proliferating pigment epithelial cells, such as vascular endothelial growth factor, fibroblast growth factor, endothelial maturity factor CD31, HMB 45, and protein S 100 [9].

Conclusion

A comparative histomorphological study of metastases and choroid hemangiomas indicates that the presence of metaplastic changes in the tapetoretinal zone, including the tumor-like proliferation of pigment epithelium, is not an accident. The permanent involvement of the tapetoretinal zone in the proliferative neoplastic and destructive process most likely indicates the leading, initiating role of these changes in choroidal angiomatosis. Similar pathogenetic relationships are already well proven for neovascularization in AMD, which occurs in the choroid, but begins in the tapetoretinal zone and is associated with the stimulating effect of angiogenesis factors of proliferating pigment cells on vascular development.

Here we first draw attention to the need for such studies, which allow us to understand the discrepancy between the morphological picture of benign angiomatosis in the choroid, and malignant, from the position of the clinic, the course, often ending with a loss of the eye.

Conflict of interest

All authors declare that they have no conflict of interest.

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References

1. Berry M, Lucas LJH. Circumscribed choroidal hemangioma: a case report and literature review. J Optom 2017;10:79-83.
2. Shields JA, Shields CL. Intraocular Tumors: An Atlas and Textbook. Publishing House Panfilova; 2018.
3. Brovkina AF. Ophthalmic oncology: a guide for doctors. M: Medicine; 2002.
4. Shanmugam PM, Ramanjulu R. Vascular tumors of the choroid and retina. Indian J Ophthalmol 2015;63:133-40.
5. Finger PT. Radiation therapy for exudative choroidal hemangioma. Indian J Ophthalmol 2019;67:579-81.
6. Reese AB, Jones IS. Hematomas under the retinal pigment epithelium. Trans Am Ophthalmol Soc 1961;59:43-79.
7. Shepkalova V, Khorosanyan-Tade A, Disler V. Tumors of the eye. M: Medicine; 1965.
8. Maftei C, Stanca HD. Intraocular ossification. Case report. Rom J Ophthalmol 2017; 61:65-8.
9. Sheiko EA. Hemangiomas in young children. International Journal of Applied and Basic Research 2015;4-2:222-8.

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