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RETINAL GLIAL TUMORS: OVERVIEW OF RARE VARIANTS

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Introduction. Typically, tumor processes in the retina in adults become objects of histological examination if they are clinically mistaken for melanomas. Since the second half of the last century in ophthalmooncology, the opinion has been established about the benign nature of glial-type retinal tumors, which in morphological terms are usually identified with astrocytomas of the brain [1-3]. Nevertheless, the clinical and morphological analysis of unique cases of retinal glial tumors makes it possible to doubt the benign nature of these neoplasms [4]. The sporadic nature of retinal glial tumors and their extremely rare occurrence in the practice of an ocular pathologist makes it difficult to systematize these neoplasms. However, such work is necessary, especially since the classification criteria used for CNS glial tumors cannot be extrapolated with respect to the retina.

Aim. The purpose of this work is to compare the clinical presentation and morphology of rare glial retinal tumors and, by their example, to discuss morphological features and classification criteria by drawing parallels with histogenetically similar brain tumors.

Material and methods. The study is based on clinical material provided by three eyeballs removed due to suspected intraocular melanoma. The surgical material was processed according to the generally accepted histological technique, followed by the preparation of serial sections from paraffin blocks. Then serial sections were stained with hematoxylin-eosin and examined under a light microscope. If possible, the material was examined by immunohistochemical analysis.

Results and discussion. Case 1. A 42-year-old patient came to the clinic due to deterioration of vision in the right eye. During the examination, vision in the right eye was less than 0.1 (not corrected), in the left eye - less than 0.3 (not corrected).Weak vision in both eyes the patient notes from his youth, when a degenerative disease of the retina was detected. The nature of the degeneration was not specified, the patient was not followed up. At the last visit, a mature cataract was found against the background of intraocular hypertension. Optical coherence tomography showed the presence of a neoplasm in the equatorial region measuring 7x1,5 mm. Due to suspicion of intraocular melanoma and difficulty in monitoring the fundus, the eye was removed. Visual examination on the sagittal section in the posterior part of the eyeball revealed the tumor-like thickening of the retina described above, but tumor growth was not detected in the choroid. Microscopic examination of the tumor revealed bundles of spindle-shaped glial cells against the background of total displacement of the typical neuronal structure of the retina (Fig. 1).

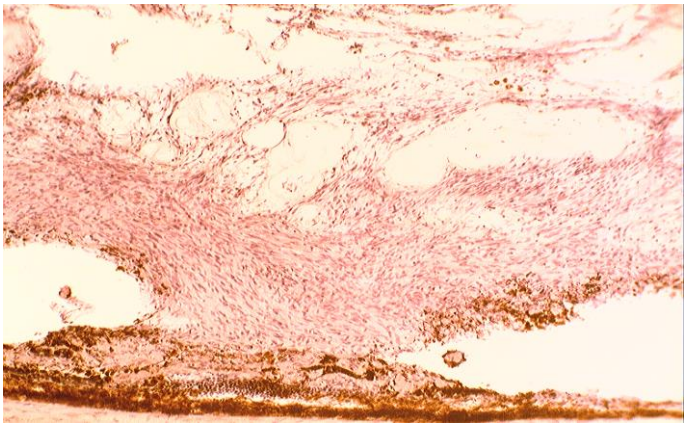

Fig. 1. Aggregates of spindle-shaped glial cells which form multidirectional bundles. Amorphous pale deposits at the top of the figure.
Stain: hematoxylin-eosin (H&E). Magnification 40 x.

In general, the tumor structure is characterized by a predominance of round-oval and elongated spindle-shaped cells with a moderately pronounced fibrous stroma (Fig. 2).The amorphous deposits noted above with tinctorial properties characteristic of glycoprotein type proteins apparently reflect the myelin-synthetic ability of tumor cells. Such patterns are rare in astrocytic brain tumors.

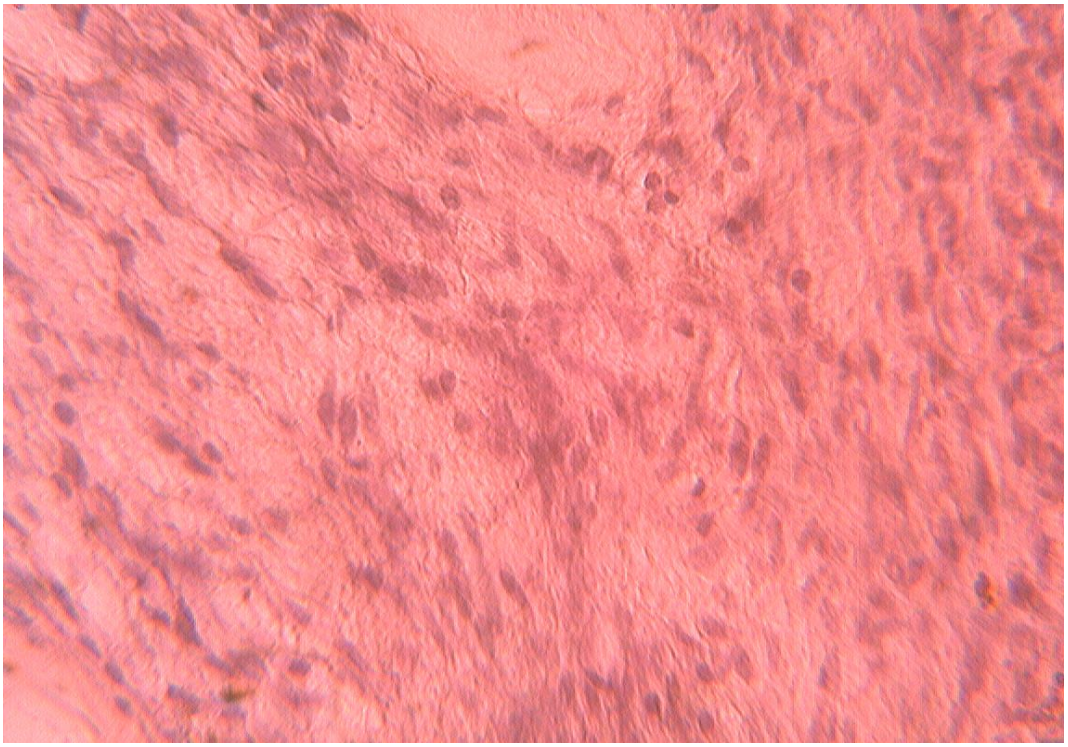

Fig. 2.Typical view of the cellular-stromal structures of the tumor. Stain: H&E. Magnification 400x.

In ocular pathology, the division of glial tumors according to the degree of malignancy is not accepted. Nevertheless, most publications emphasize the benign nature of ocular astrocytomas, although the arguments presented cannot be considered convincing. As a rule, the authors rely on the absence of pronounced cellular polymorphism, atypical cells, and mitoses. However, these criteria for tumors of this histogenetic group are not as indicative as, for example, for epithelial carcinomas. On the other hand, regarding the above case it is worth paying attention to a sharp violation of the typical histoarchitectonics and complete erasure of the border between the tumor tissue and the surrounding retina. At least, this corresponds to grade 2-3 malignancy in brain tumors.

Case 2. An intraocular tumor in a 64-year-old woman was verified as Grade-2

glioblastoma based on immunohistochemical data. However, despite the use of the same markers for immunohistochemical analysis of tumors of this histogenetic group, we cannot transfer the WHO [5] classification criteria adapted for brain tumors to retinal neoplasms. So, brain glioblastoma of the 2nd degree of malignancy can invade the surrounding tissues, but does not metastasize within the brain, and even more so outside it. In our case, the tumor not only filled the entire eye cavity, displacing normal retinal elements, but also formed a massive (2x3 cm) metastasis in the orbit. This clinical observation refutes the idea of benign retinal glial tumors. The histomorphological picture of this neoplasm has many common patterns with the previous one, while there is no polymorphism and cellular atypism characteristic of glioblastoma multiforme, the most malignant brain tumors (Fig. 3-4).

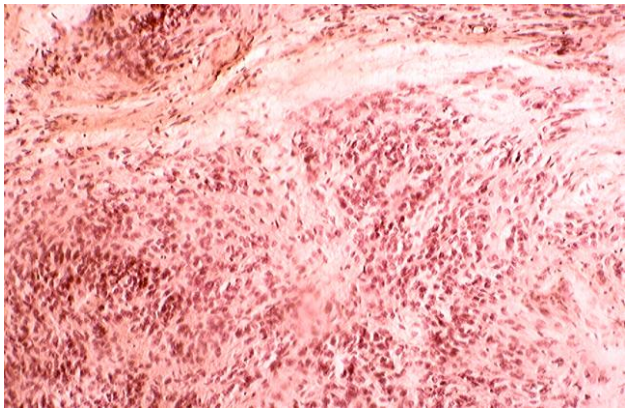

Fig. 3. Characteristic patterns of retinal glioblastoma of the 2nd degree of malignancy. Stain: H&E. Magnification 200x.

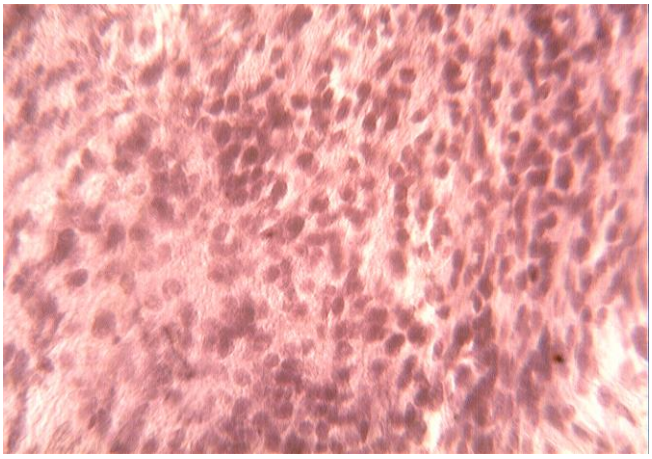

Fig. 4. Cytological details of retinal glioblastoma of the 2nd degree of malignancy: moderate cellular polymorphism without noticeable cellular atypia. Stain: H&E. Magnification 600x.

Case 3. Patient 44 years old. A grayish-white tumor node, 5x3 mm in size, was found in a subatrophic eye as an accidental finding. The neoplasm was located in the peripapillary zone of the posterior part of the eye. Microscopic examination in the area of tumor growth revealed a total violation of the typical neuronal stratification of the retina with its replacement by glial tissue with a predominance of fibrous stroma with a small number of cellular elements against the background of neovascularization (Fig. 5-6).

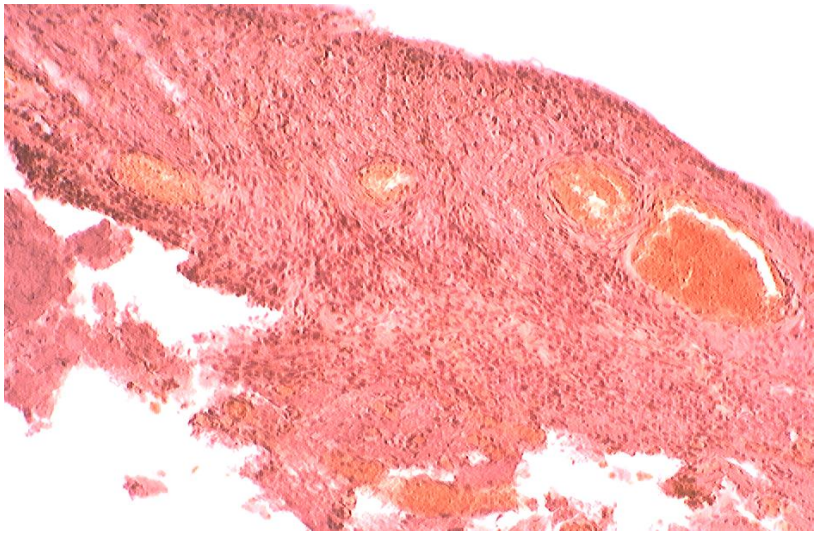

Fig. 5. Overview of a glial tumor of the retina. Total replacement of the structure of the retina in the area of tumor growth.
Stain: H&E. Magnification 40 x.

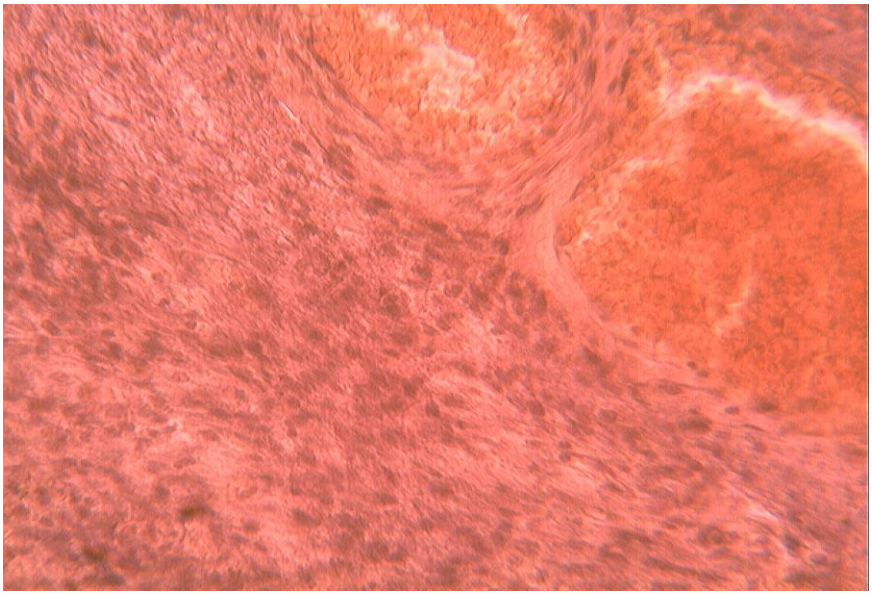

Fig 6. Cytological details of a glial tumor. Stain: H&E. Magnification 400 x.

Conclusion. Comparison of the histological picture of three rare retinal glial tumors shows the relative monomorphism of their cellular composition in the absence of mitotic patterns and atypical monster cells which are characteristic for malignant variants of brain glioblastomas. Similar histological patterns can sometimes be found in detached retina as a manifestation of post-traumatic gliomatous transformation. Thus, anamnestic data (absence of trauma) and the detection of localized tumor growth was often the basis for the diagnosis, although, as in cases 1 and 3, the latter may be poorly noticeable. In all cases we observed, degenerative changes in the retina and subatrophy of the eye occurred regardless of the size of the tumor. All this gives grounds for early removal of the eye (when organ-preserving therapy is not possible), since in an advanced case, a retinal glial tumor can behave no less aggressively than intraocular melanomas.

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