EUROPEAN OPHTHALMIC PATHOLOGY SOCIETY

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Material distributed **glass slide**

Title: Focal Reactive Nodular Retinal Gliosis

Clinical & Personal History: The patient is a 64 yo men with low vision in both eyes (amblyopia right eye-OD-/ chorioretinal scar since childhood left eye –OS-) with sudden vision loss in the OS.

Ocular examination showed best corrected visual acuity (VA) 0,16 in the OD and light perception in the OS. OD was unremarkable. Under slit lamp examination OS show posterior synechiae, iris rubeosis cataract.Fundus eye examination exhibit vitreous haze and hemorrhage in reabsorption with suspected inferior mass. Ultrasound demonstratedan infero-temporal tumor, mushroom shaped measuring 10.4 x 8.2x 3.3 mm. The mass was hyperintense in T1 and hypointense in T2-weighed magnetic resonance images consistent with melanoma. Systemic extension test were negative for metastasis. Enucleation was performed on the basis of intraocular tumor consistent with melanoma with 2ª vitreous hemorrhage and inflammatory signs.

Ocular Pathology.

Gross examination: Left eye measuring 24x24x23 mm. No signs of extraocular extension were observed. A nodular amelanotic mass was observed involving less than one quadrant ofretina at equatorwith respect of underlying choroid and blurred overlying vitreous.**Microscopic Description**: A well delimited nodular retinal proliferation was observed composed of fusiform cells, arranged in bundles with round or oval nucleus without signs of atypia. Numerous blood vessels with thickened and hyalinized wall were present and hemorrhage foci. Adjacent retina shows degeneration and intrarretinal exudates. Immunohistochemical stains showed intense positivity for GFAP but not for S 100. Ki 67 revealed <5% of proliferative activity.

Final Diagnosis: Focal reactive nodular gliosis overlying chorioretinal scar

DISCUSSION

Retinal glial tumors of the retina can be classified into astrocytic hamartoma, acquired retinal astrocytoma and nodular/ massive retinal gliosis. Each has different clinical manifestations.

Astrocytic hamartoma is a congenital tumor, usually seen in patients who have some manifestations of tuberous sclerosis complex(TSC- sebaceous adenoma- Bourneville Disease. Ophthalmoscopy usually appears as a grayish-yellow mass on the retina that may show calcification.Although it is generally a stationary lesion, can lead to exudative retinopathy, retinal detachment and neovascular glaucoma. Histopathologically it is composed of well differentiated fusiform glial cells.

Acquired astrocytoma general occurs in older individuals who do not have TSC. It presents as a solitary mass in the neurosensory retina, yellowish, with a tendency to slow and progressive growth. It can cause intraretinal exudation and retinal detachment, as well as vitreous hemorrhages. Histopathologically, it is composed of mature glial cells similar to those of astrocytic hamartoma, without calcification.

Nodular and massive reactive retinal gliosisincludes a spectrum of reactive proliferations of glial retinal cells surrounding blood vessels, ranging in size from small nodules to massive lesions that mayfill the eye, thus being called massive retinal gliosis. These lesions are reactive in nature, arising from retinal astrocytes or Müller radial glial cells.It usually appears in eyes with chronic inflammatory pathologies and often occurs in blind eyes.Clinically the lesion may be asymptomatic and incidentally discovered during fundus examination, or it may cause decreased vision, primarily due to exudative retinal detachment or exudation

Histopathologically, it consists of a dense mass of well-differentiated astrocytes and highly vascularized.The lesion is composed of spindle-shaped cells and may contain hair-like (pilloid) processes or Rosenthal fibres. There are often scattered sclerotic vascular channels and capillaries within the lesion. Subretinal proteinaceous exudates may extend beyond its borders. Fibrous (or even osseous) metaplasia of the RPE at the base of the lesion can be prominent.The astrocytic cells are immunopositive for GFAP and S100 protein; mitotic figures are absent or rare, and Ki 67 index is <5%. Differential diagnosis includes pilocytic astrocytoma, diffuse astrocytoma, giant cell astrocytoma, astrocytic hamartoma (phakoma) and retinal haemangioblastoma.

Jakobiec et al (2016) in reviewing all published cases were able to stablish 3 subgroups: **focal nodular gliosis, submassive gliosis and massive gliosis.** All lesions were composed of mitotically quiet, compact spindle fibrous astrocytes. Histopatologically all 3 tumoral categories were accompanied by progressively fibrous and osseous metaplasia of the Pigment epithelium, micro/macrocysts, vascular sclerosis, exudates calcospherites and Rosenthal fibers among the proliferating astrocytes. Immunohistochemistry was positive for GFAP, nestin in most. The nonneoplastic nature of all categories was confirmed by absent TP53 expression, Ki-67 negativity and intact p 16 expression. These findings indicate an intrinsic attempt to regulate and maintain a low level of glial cell proliferation. These categories appeared to constitute a spectrum: focal nodular tumors encompass lesions previously called retinal vasoproliferative lesions, which display the same histopathologic and immunohistochemical findings as the 3 categories of retinal gliosis described.

Prognosis and treatment. The prognosis for focal reactive (nodular) gliosis is uncertain. Vision maybe threatened by either vascular exudation or retinal detachment, and the target of the treatments are the blood vessels in order to decrease exudation.The disease may require treatments such a laser photocoagulation, cryotherapy, plaque brachytherapy, antiangiogenic therapy, vitreoretinal surgery or enucleation.

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