

Retinal Primary Benign Tumor with Unusual Histopathological Exam

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Abstract

Purpose: To describe a new subtype of retinal tumor.

Conclusion: To the authors' knowledge, we report a case of benign retinal tumor not previously described, that seemed a vasoproliferative tumor but with unusual histopathological markers.

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Dear Editor,

Retinal tumors have different clinical and histopathological characteristics that allow its classification in different groups. However, herein we report a case of a primary benign tumor that clinically seemed a vasoproliferative lineage, but with unusual histopathological staining exam.

The patient (70-year-old man) attended our department complaining of blurred vision in its Right Eye (RE) that had begun eight months ago. No more diseases were documented. Best-corrected visual acuity was hand motion in its RE and 20/30 in left eye. On dilated funduscopy and ultrasonographic examination of RE, an exudative retinal detachment with macular involvement and a sub retinal mass could be detected.

Criotherapy in the base and the borders of the tumor was performed, solving retinal detachment.

A vasoproliferative tumor was then appreciated, showing fast angiofluorescein staining (Figure A and B) with a thickness of 13 to 11 mm, and medium to low internal reflectivity. Endophitic growth, no sub retinal fluid and medium-to-high internal reflectivity could be observed in ecografic exam. MRI was described as a well-defined and homogeneous tumor that seemed hemangioma, without involvement of the rest of the orbit. As the characteristics seemed a benign tumor, the patient was followed up during 10 years, when it started with ocular pain and high intraocular pressure (29 mm Hg in treatment with fixed combination of brimonidine 0.2 % and timolol 0.5 %). The patient chose to underwent an enucleation. On pathological examination the specimen showed micro vacuolated polygonal cells with broad cytoplasm and rounded nuclei (Figure C). A fine and branched vascular structure was distributed among the cell groups (Figure D). No mitosis was detected, and the rate of proliferation was less than 5%. No areas of necrosis were observed. Immunohistochemistry analysis was positive to CD99 (Figure E), inhibin (Figure F), and vimentin (Figure G). Other tested markers were negative such as melanocytic (s-100, HMB45 and MELAN-A), epithelial (CKAE1/AE3, CK8 and CAM 5.2), muscle (actine), vascular (CD34 y CD 31) and neuroendocrine (cromogranine, synaptophysin and enolase) markers. After a follow-up of 20 months, there were no signs of metastases or local recurrence.

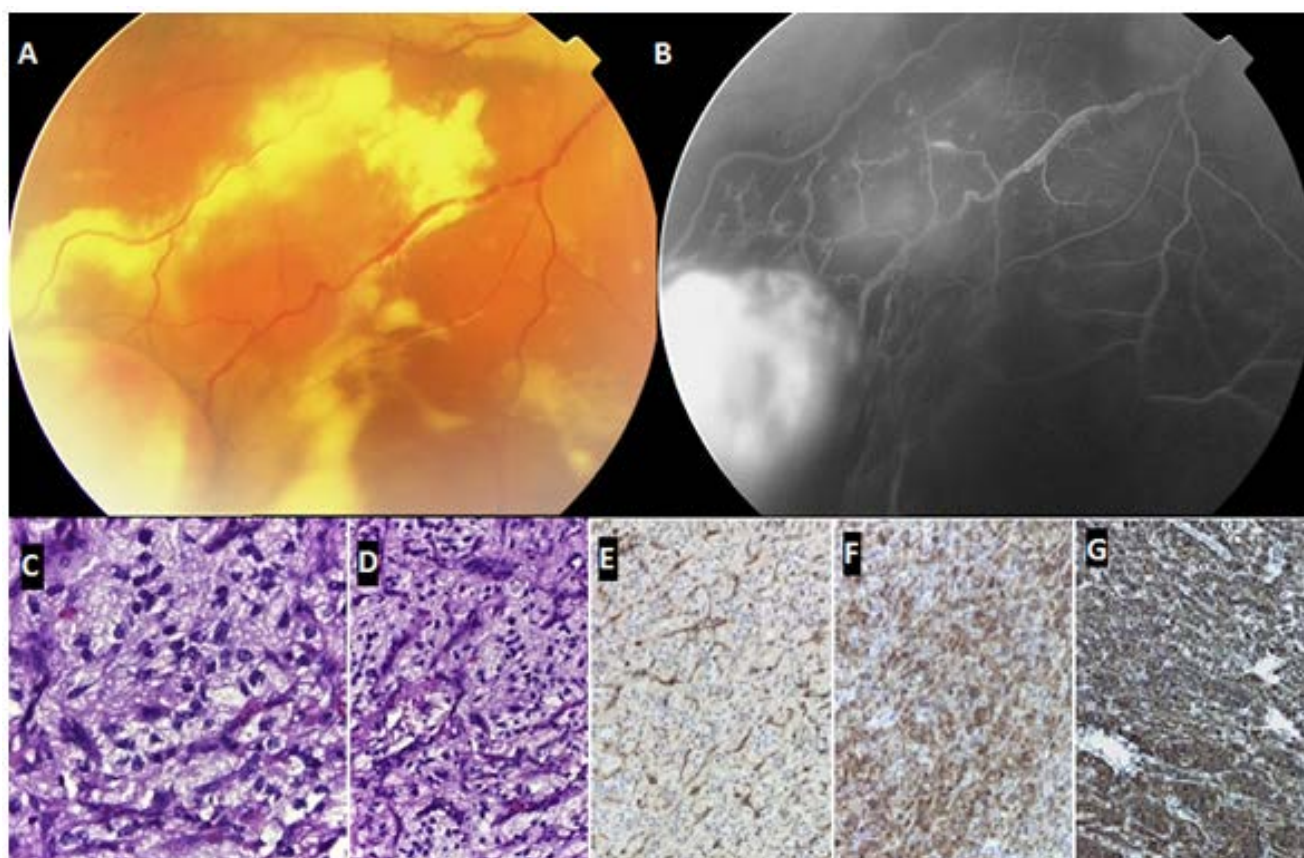


Figure: Funduscopy image shows a peripheral located tumor with intraretinal exudation (A). Angiofluorescein image shows fast staining (B). Different areas of cytologically micro vacuolated polygonal cells (C, D) (hematoxylin-eosin). Positive staining to CD99 (E), inhibin (F), and vimentin (G) (original magnification $\times 200$).

Clinical characteristics of this tumor remind a retinal vasoproliferative tumor, which usually presents as a vascular mass located in the periphery of the retina, near the ora serrate. Most affected group of patients is middle and old-aged. This benign tumor can produce intraretinal and sub retinal exudation, sub retinal fluid, and cystoids macular edema [1]. Clinical course of these tumors is variable. Some cases are stable for years whereas others grow and may be complicated by exudative macular changes or exudative retinal detachment [2]. Clinical differential diagnosis of vasoproliferative tumors should be set with capillary hemangiomas of von Hippel-Lindau disease. This disease usually presents with multiple angiomas,

in young patients, and with marked dilated afferent and efferent vessels. In addition, there is a family history and associated kidney or cerebellum tumors are often found.

However, histopathological exam of this tumor revealed some unusual markers not associated with vasoproliferative tumors [3]. This tumor showed a positive result to CD99, inhibin and vimentin.

Combination of vimentin and CD 99 markers is associated with Ewing Sarcoma, and it has already been published a case of a tumor of this lineage located in retina [4]. Nevertheless, Ewing Sarcoma is usually positive to other markers as enolase and synaptophysin [4] (negative for the reported tumor), and negative to inhibin. Besides, reported retinal Ewing Sarcoma was found in an 11 year-old child, and with a white appearance [4], whereas tumor herein described was found in a 70 year-old man with an orange color. On the other hand, it is published a case of von Hippel-Lindau Disease with angiomas located in retina that were positive to inhibin and enolase, but negative to other markers [5].

To the authors' knowledge, we report a case of benign retinal tumor not previously described, that seemed a vasoproliferative tumor but with unusual histopathological markers.

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