Recurrent Hyphema in an Iris Tumor, a Case Study and Review of Literature.

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Abstract: A young male presented to the ophthalmology department with history of recurrent bleeds into the left eye. He was thoroughly evaluated, and surgically managed, visually rehabilitated. The literature on tumors of the iris is reviewed and protocols for management of iris tumor with recurrent hyphema are discussed. No financial disclosures. The authors acknowledge the evaluation performed at SankaraNethralaya, Chennai and the histological examination and report by Anand Institute of Laboratory Sciences, Bengaluru.

Keywords: Recurrent hyphema, Iris hemangioma, iris tumor

I. Introduction

Hyphema in an young adult male is usually due to trauma, or rubeosis (following a retinal vein occlusion/perivasculitis retinae / diabetic retinopathy), tumors of the iris, sickle cell trait, chronic uveitis, anticoagulant overdose (International normalised ratio >5), idiopathic thrombocytopenic purpura or hemophilia. Recurrent hyphema is difficult to treat, hence it is important to find the cause and manage it before the onset of blinding complications. It is essential to reduce the intraocular pressure in order to avoid blood staining of the cornea and compromise of optic nerve function. However timely surgical evacuation of the hyphema is the only definite means of reducing the rate of complications in the long term.

It is important to rule out melanoma of the iris in any tumor of the iris. Iris tumors have to be evaluated using ultrasound biomicroscopy and anterior segment optical coherence imaging.

Tomography (OCT) to determine the extent of involvement and plan surgical management. The case proves that vision can be saved by surgical intervention despite of large hyphemas recurring over a course of years.

II. Case report

History: A 32 year old male presented to ophthalmology department, with complaints of pain, redness and blood in the left eye for the last few months. The complaints began one year back with redness, pain and loss of vision for which he visited a tertiary eye care center and had some investigations performed. He was treated with topical medication and the symptoms were temporarily relieved. However the symptoms recurred within a few months and the patient was told to use the same medication and wait for improvement.

On examination

He was noted to have an unremarkable RE (vision, anterior and posterior segment were normal). LE revealed hypopigmented lobulated iris mass on the superior pupillary border with hemorrhages on the surface, hyphema measuring 5mm in the anterior chamber, Figure 1. Fundus evaluation showed a normal disc & posterior pole. Peripheral fundus details were not clear. His vision LE was reduced to 6/24P, N10. Intraocular pressure was RE14 mmHg and LE 25mmHg, applanation on topical antiglaucoma medication.

III. Ancillary Investigations

Complete blood count revealed eosinophilia (N 59 L 22 M3, E14, B 1) and absolute eosinophil count 1.80 cubic microlit (Normal 0.4) but normal platelet count and hemoglobin levels. Bleeding time, clotting time were unremarkable as was E.S.R(10mm), the blood sugar and lipid profile. Ultrasound biomicroscopy done (06/07/2012) showed ...3

LE ruptured iris cyst seen 10-2 clock hours Limiting to anterior lens capsule, lens in situ, homogenous reflective echoes hyphemanoted in the anterior chamber. Possibility of anterior capsular rupture at 3-5 clock hours, ciliary body normal, Figure 2.

B Scan ultrasound LE was normal.
MRI brain (MRI number 2761 14/11/2012) was performed, and ultrasound of the abdomen (15/11/2012) was performed to rule out visceral angiomatosis. The reports were unremarkable.

An anterior chamber paracentesis was performed after informed consent, to look for malignant cells in the hyphema, but histopathological examination, after centrifugation of the anterior chamber aspirate did not reveal any significant findings.

IV. Diagnosis and Treatment

There was no improvement in the hyphema. The patient was then informed of the need to excise the tumor with possibility of recurrence of bleed, cataract formation and visual loss in vernacular, and consent for the procedure obtained. The patient underwent LE sectoral iridectomy and hyphema evacuation under local anesthesia. The iris was sent for histological examination to two different labs. Postoperatively the LE showed cataractous changes and visual acuity deteriorated to only counting fingers on first postoperative day due to the intumesence caused by loss of capsular integrity in the process of iris mass excision. This was anticipated as the mass adherent to the lens on ultrasound biomicroscopy, it also showed loss of capsular integrity at 3-5 clock hours. The lens was removed by lens aspiration and posterior chamber IOL implantation was performed under local anesthesia. ...

Postoperative visual acuity eventually improved to >6/12 with -1.0DCy 90 degrees. NV N8 with best correction.

Figure 3The report from the college was nonspecific, but the report from the private lab was Cavernous hemangioma of the iris. Figure 4

V. Discussion

Vascular tumors of the iris are classified as Iris capillary hemangiomas, iris cavernous hemangiomas, iris microhemangiomas, iris arteriovenous malformations, and iris varices, (1). These simulate iris melanomas. Factors predictive of iris nevus growth to melanoma include - age (<40 years at presentation), recurrent hyphema, 4:00 to 9:00 clock hour location of tumor, tumor involving entire iris surface, ectropionuveae, and feathery tumor margins. Additional factors include tumor seeding in the anterior chamber angle, feeder vessels, and nodule formation. These factors can be remembered using the mnemonic ABCDEF, representing A - age, B - blood, C - clock hour inferior, D - diffuse, E - ectropion, and F - feathery margin. (2)...5

Signs of malignancy are ectropion, secondary cataract, secondary glaucoma, hyphema. (3)

The iris melanoma can cause heterochromia iridis. Rarely, the iris melanomas can produce lightly pigmented nodules - tapioca melanoma which cause glaucoma following invasion of the angle of the anterior chamber. (4) Careful documentation and histological examination is suggested even in the absence of evident tumor growth, to rule out malignancy as 4-5% of suspicious iris nevi transformed to malignant melanomas. (4, 5) When hyphema is disabling and vision threatened, tumors are best treated by sectoral iridectomy. Melanomas of the iris treated thus do not recur. (6)

Transient hyphemawas the commonest mode of presentation in vascular tumors of the iris. (7) The commonest vascular tumor of the iris was racemose angioma, followed by cavernous angiomas, capillary hemangiomas, varices and microhaemangiomatosis. Most vascular tumors of the iris are confined to the iris except cavernous hemangiomas which show systemic involvement. (8)

Anterior uveal metastases must be considered in the differential diagnosis of the iris tumors. They present as yellowish pink nodules and are mostly seen at the lateral and medial parts of the iris on account of the long posterior ciliary arteries. (9)

VI. Conclusions:

Traditional teaching is to observe hyphema caused by an iris tumor, however, evaluation by ultrasound biomicroscopy and anterior segment optical coherence tomography is essential. These techniques are able to determine the depth of tumor invasion or cyst extension with a resolution of 5-10 microns. These determine the future course of action. In diagnosis of any tumor, it is imperative to send the biopsy to two independent, pathology departments to confirm the diagnosis. ……7

References

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Figure 1 Iris tumor with hyphema

Figure 2 Ultrasound biomicroscopy – Ruptured Iris cyst

Figure 3 Postoperative view – Complete iridectomy, cataract extraction, intraocular lens in situ
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Figure 4 Histopathology Cavernous hemangioma of the iris