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Case Report

Retinoblastoma presenting as total hyphema: Three year follow-up

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Abstract

Retinoblastoma very rarely presents as total hyphema. Our patient presented at an early age of 7 months. Follow-up of 3 years shows that unilateral group E retinoblastoma was treated successfully with enucleation and adjuvant chemotherapy. The fellow eye remained normal during this period. The factors associated with delay in treatment are also described. Reports like the present case add to the information available about advanced staging of retinoblastoma at the time of presentation, seen in cases with spontaneous hyphema due to the tumor.

Keywords: Retinoblastoma, Hyphema, Enucleation

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Introduction

Hyphema is considered among the high risk clinical predictors in retinoblastoma. It may occur spontaneously or after a history of trauma. Long term follow-up is required after treating the patient to monitor the course of the disease. We describe the management and follow-up of a patient presenting with total hyphema. He had anteriorly located tumour mass on B-scan of the left eye which was also confirmed on imaging of the orbits and brain.

Case report

A 7-month-old patient presented to our tertiary care eye hospital with complaints of discoloration of the left eye for the last two months. There was no history of trauma or previous ocular surgery. On clinical examination in the office and examination under anesthesia (EUA) the right eye had a normal anterior segment. Gonioscopy showed normal angle structures in the right eye. Dilated fundus examination with indentation was also within normal limits. Total hyphema was noticed in the left eye obscuring any posterior segment examination. Intra ocular pressure was 12 mmHg in the right whereas the left eye was soft. B-scan of the left eye showed hyper echoic shadow in the anterior vitreous (Fig. 1). B-scan also showed anteriorly located calcification in the post segment, apparently not attached to the retina. Diffuse echogenicity was also seen in the vitreous cavity.

Imaging of the brain and orbits showed increased attenuation of fluid in the anterior and posterior chamber of the left eye presumably due to hyphema. The vitreous was also of high attenuation with an area of coarse calcification measuring 9 mm associated with a suspicious intraocular mass in relation to the chorioretinal layer involving up to 20% of the retinal surface. There was no extra-ocular extension noted in the left orbit. Right orbit and its contents were normal. Topical steroids and cycloplegic eye drops were prescribed. The parents were meanwhile counseled for the possibility of

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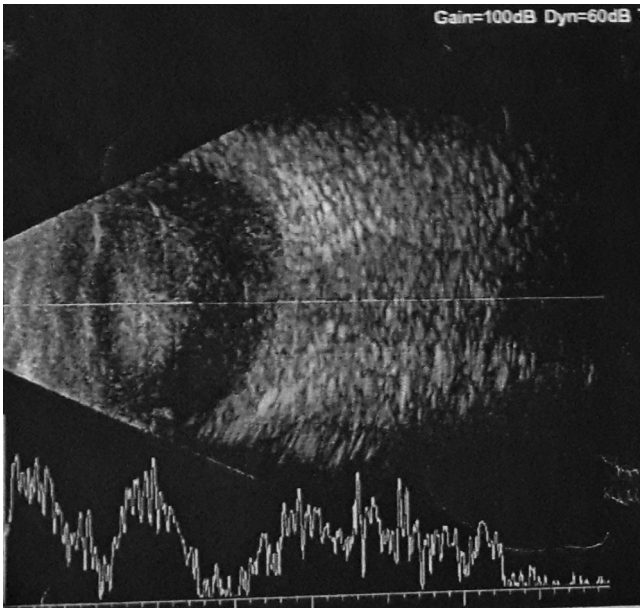

Fig. 1. B-scan of left eye.

enucleation of the left eye considering the calcification in the intraocular mass on imaging. Bleeding profile of the patient was within normal limits. There were no signs of non-accidental trauma.

The patient missed follow-up visit and took 1 month to return probably due to the enucleation being a hard decision for the parents. The hyphema in the left eye had cleared to some extent by this time (Fig. 2). The eye was firm this time. Suspicious white deposits on the iris root were seen in the anterior segment. A white lesion just behind the iris was visible. Left enucleation with implant was carried out for a group E tumor.

Histopathology revealed multifocal Retinoblastoma. Flexner Wintersteiner rosettes were identified. Dystrophic calcification and necrosis were also present. Tumor involved the anterior chamber and angle, as well as the ciliary body and iris. Focal choroidal invasion was noted. There was no optic nerve invasion and the resection margin of optic nerve was not involved. Vitreous hemorrhage was present. TNM pathological staging was pT_2 N_x M_x. The patient was sent for chemotherapy with Carboplatin, Etoposide and Vincristine (CEV protocol) considering the risk factors on histopathology. Regular EUAs were carried out during and after completion of chemotherapy. Follow-up MRI shows no definite evidence of residual or recurrent disease. Last EUA done after 3 years of initial diagnosis showed a normal right fundus examination and the prosthetic eye was reinserted after noting a normal left socket.

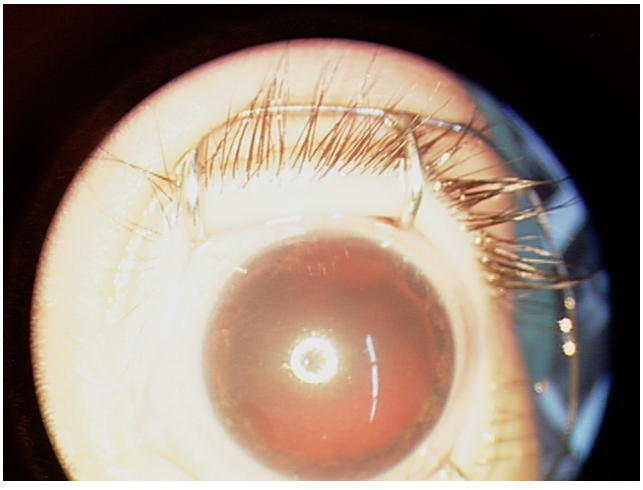

Fig. 2. Clearing hyphema.

Discussion

The first case of retinoblastoma presenting as total hyphema was reported in 1985.[1] Balasubramanya reported hyphema in only 0.25% of patients in a series of 392 cases of retinoblastoma. [2] Hyphema as a presenting feature was seen in 2 (5%) cases in a study of 57 eyes of 40 patients.[3] Similarly a review article described hyphema as a presenting clinical feature in 1% of retinoblastoma cases.[4] The patients presenting as total hyphema and later confirmed as having retinoblastoma were generally older than our patient.[1,5] Total grade-4 hyphema and raised intraocular pressure were the presenting features of unilateral retinoblastoma in a two-year-old female.[5] Biopsy revealed the presence of Wintersteiner rosettes as was found in our case. History of trauma is reported in many of the cases described in the literature. Every effort should be made to exclude a tumor in all cases of hyphema, even in cases of well documented ocular trauma.[6] Our patient did not have any history of trauma and the age group was more of non-accidental injury, if trauma was to be suspected at all. However, no such signs were present in our patient.

Our patient had total clotted blood in the anterior chamber. The hyphema was grade IV, often referred to as 8-ball hyphema. Since tumor growth is dependent on vascular supply, retinoblastoma can lead to development of hyphema. The relatively anterior location of the tumor which apparently did not seem to be attached to the retina on B scan and CT scan created suspicion about retinoblastoma initially, especially when the visibility was obscured due to the total hyphema. The presence of calcification on imaging and suspicious anterior segment deposits was seen on follow-up. These findings were the deciding factors about counseling for enucleation to confirm the diagnosis. Precise evaluation of such cases may be done by ultrasound biomicroscope (UBM).[7]

Atypical presentations of retinoblastoma usually present with advanced disease.[2,5] All patients of retinoblastoma presenting with atypical features required enucleation with adjuvant chemotherapy or radiotherapy or both.[2] The 4 year old child reported with history of trauma also had enucleation and chemotherapy after diagnosis of retinoblastoma.[6] Our patient also had adjuvant chemotherapy for the unilateral retinoblastoma after enucleation due to the risk factors noted on histopathology. In contrast a 2 year old child with hyphema and iris deposits similar to our patient, despite having high risk factors on histopathology of the enucleated eye did not receive any further treatment and was doing well at 5 years of age.[8] In the authors' practice the patients with histopathological risk factors are referred for adjuvant chemotherapy. Drainage of anterior chamber should be avoided as it may result in metastases with an untoward

out come as reported in a 6 year old girl.[9] Patients with hyphema should avoid paracentesis till retinoblastoma is excluded.[2] A study of enucleated eyes of retinoblastoma patients showed higher incidence of high risk histopathology findings in associated clinical features of glaucoma and neovascularization of iris. Hyphema was also included in clinical variables strongly associated with high risk histopathology findings in this study.[10] Our patient had focal choroidal invasion (less than 3 mm involvement of the choroid that does not reach the sclera) which is not included in high risk histopathology factors. The risk factors encountered in our patient were the presence of tumor in the anterior chamber and angle, as well as the ciliary body and iris. Three years after enucleation and completion of chemotherapy our patient has not shown any signs of recurrence or metastatic disease on EUA and repeat MRI of orbits and brain. The patient is being followed for any late appearance of bilaterality of the disease as has been reported after a long interval at the age of 9 years.[11] Atypical presentations can delay diagnosis and worsen prognosis in cases of tumors.[5] Spontaneous hyphema is included in the list of atypical presentations of retinoblastoma.[4] Ophthalmologists should apply all non-invasive diagnostic measures to reach a definite diagnosis and treat the disease as soon as possible. It is also important to integrate counselors in managing such patients. A study reported that parental knowledge of retinoblastoma nature and heritability is essential for good patient outcomes.[12] Translating this knowledge into appropriate screening of at-risk children however is still deficient according to the above study.[12] Similarly it is important to develop a means of tracing defaulters as enucleating the eye without further monitoring may not achieve the desired treatment goal.

Conflict of interest

The authors declared that there is no conflict of interest.

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