HYPHEMA REVEALING RETINOBLASTOMA IN CHILDHOOD. A CASE REPORT.

CHRAIBI F, BHALLIL S*, BENATIYA I*, TAHRI H**

ABSTRACT

We present a case dealing with an uncommon presentation of retinoblastoma. An 4- year-old boy presented to the ophthalmic department for a red painful eye following trauma. The examination showed decreased visual acuity, total hyphema and ocular hypertony. Ocular ultrasonography revealed an intraocular process. CT-scan of the orbit was consistent with a retinoblastoma. Treatment consisted of an enucleation and chemotherapy. This paper stresses the fact that presentation of retinoblastoma is not stereotypic. Every effort should be made to exclude a tumoral intraocular process in all cases of hyphema, even in cases of well documented ocular trauma.

KEY WORDS

Retinoblastoma, hyphema, ocular trauma, ultrasonography

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* *University Hospital Hassan II of Fez*

Submitted: Apr 01, 2009
Accepted: July 20, 2011

INTRODUCTION

Retinoblastoma is the most frequent primary ocular malignant tumour in childhood. Clinically, it presents itself most often as a leucokoria and a strabismus (1). In this paper, we present a less common clinical pattern of a retinoblastoma mascarading as a hyphema.

CASE REPORT

Our patient was a 3 year-old boy with no particular past medical history. Chief complaint was a left painful red eye coexisting with a decreased visual acuity following an unimportant eye trauma.
On presentation, his visual acuity was 20/20 in the right eye (RE) and was reduced to bare-light-perception in the left eye (LE). Anterior segment and fundus examination in the RE were unremarkable. In the LE, the examination revealed a corneal abrasion, a 8 millimiters height ball hyphema, and an elevated intraocular pressure to 50 mmHg. Ocular ultrasonography of the LE revealed an intraocular mass with hyperechogenic features. Further radiologic exploration consisting in an orbital and cerebral CT-scan showed intratumoral calcifications that were very suggestive of a retinoblastoma (*Figure 1*).
Screening for local and distant metastasis showed no evidence of tumoral extension. The left eye was enucleated and replaced by an ocular prothesis. No extraocular extension of the mass was noted.
Histopathologic analysis of the enucleated eye confirmed the diagnosis of retinoblastoma and showed massive choroidal invasion, anterior chamber involvement and iris invasion. The optic nerve was not involved. The patient received three courses of adjuvant chemotherapy associating Cyclophosphamide (300 mg/m^2), Doxorubicin (60 mg/m^2), and Oncovin (1,5 mg/m^2).
At two years post-treatment, there was no evidence of extraocular extension or distant metastasis.

DISCUSSION

Classical clinical findings of retinoblastoma include leucokoria and strabismus. In advanced cases, retinoblasotma may be revealed by a proptosis associated with orbital inflammation simulating an orbital cellulitis or even more by a spontaneous hyphema.

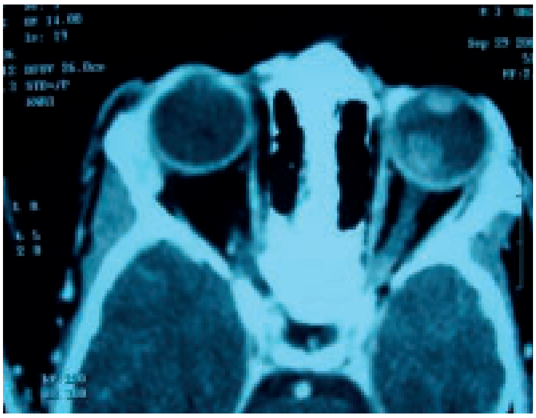

Fig. 1: orbital CT-Scan confirming intraocular calcifications in the left eye suggestive of retinoblastoma.

As a similar case was reported by Humerto et al in 2007, we elected to emphasize this relatively uncommon but not unusual form of initial presentation of an advanced retinoblastoma (2).

Spontaneous bleeding in the anterior chamber is suspected to originate from iris neovascular vessels (3). In a series of 392 cases of retinoblastoma presented by Balasubramanya et al, hyphema only represented 0.25% of all clinical presentations (4). Moreover, atypical presentations of retinoblastoma are usually associated with advanced disease as been found by Shields et al. and they are also associated according to Kashyap et al. with a higher incidence of high risk histopathology findings (5, 6).

Anyway a spontaneous hyphema or a hyphema following an insignificant ocular trauma in a child must be handled with special care. Any operative management (i.e hyphema drainage) is strictly contra-indicated until a diagnosis of a malignancy could be ruled out by adequate investigations using at least ocular ultrasonography. This may be challenging is some cases. In a case report described by Murthy et al., hyphema drainage was performed in a 6-year-old girl with a total hyphema following insignificant trauma while the ocular ultrasonography was consistent with a vitreous hemorrhage (7). Postoperative course was complicated by the development of a vascular conjunctival mass and massive cervical lymphadenopathies. The patient died 7 months later. Histopathological postmortem investigations pointed to a diagnosis of metastatic retinoblastoma.

CONCLUSION

Especially in childhood, it is crucial to rule out a intraocular tumoral process in any case of hyphema precluding the eye fundus examination. This also concerns *a fortiori* all cases of well documented ocular trauma. Ocular ultrasonography is very useful in managing these cases. Orbital CT-scan should be performed if ultrasonography is not totally reliable, especially in cases of vitreous hemorrhage.

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Correspondence:
Dr FOUAD CHRAIBI
University Hospital Hassan II of Fez,
Omar Drissi Hospital
Department of Ophthalmology,
Batha, Fez, Morocco.
Email: fouadchraibi@gmail.com
Telephone number: +21261328182
Fax number: 21235634551