lntra- and extraconicalcavernoushemangiomas in children: the difficulty of therapeutic management

Abstract

This article describes the case of a 10-year-old girl with a rare presentation of extra- and intra-conical cavernoushemangioma in children. Afterundergoingcompletesurgical excision, the exophthalmoscompletelyregressed, and no recurrencewasobservedduring the two-year follow-up period. The treatment options discussedincludedintratumoralcorticosteroid injection, oral corticosteroidtherapy, and surgical excision.

Keywords : hemangioma,children,cavernous

Introduction

Cavernoushemangiomas are benigntumorsthatprimarily affect the orbit and are more commonlyfound in youngadults. However, they are quite rare in children. In children, capillaryhemangiomas are the more prevalent type of vasculartumors. Thesetumors are usuallyfound on one side and can differ in both location and size. Surgical intervention istypicallyrecommended for treatment, but it can bequitechallenging due to the tumor's location and itsproximity to important vascular and nerve structures within the orbit.

Wepresent a unique and uncommon case of a 10-year-old girl whowasdiagnosedwith an extra- and intra-conical cavernoushemangioma.

Case report

The case involved a 10-year-old girl with no significantmedicalhistory. At the age of 6, shedeveloped progressive unilateralleftexophthalmos, alongwith a bright-red mass in the medial canthus. There were no otherassociatedsigns. Ophthalmologicalexaminationrevealed an irreducible, painless, non-pulsating, and non-puffingleft axial exophthalmoswithoutanysigns of inflammation and a brightpink-red lesion in the medial canthus (figure 1). Her visualacuitywasestimated at 9/10 in the lefteye and 10/10 in the right eye. Her ocularmotilitywaspreserved, and fundus examinationshowed no abnormalities. An MRI wasperformed, whichrevealed a left orbital lesional process. The lesion

appearedheterogeneous on the MRI, withisosignal T1 and hypersignal T2. It showedmoderatecontrastenhancement and measured 44 mm anteroposteriorly, 30 mm in width, and 36 mm in height. The lesion infiltrated the intra- and extraconical fat and involved the optic nerve, although the nerve remainedwellindividualized. The lesion predominantlyaffected the internal angle of the orbit, resulting in grade III exophthalmos (figure 2). The patient wasreferred to the neurosurgicaldepartment for management. She underwent complete en bloc excision of the leftintraorbital lesion using a high endocranialapproach. The lesion wasencapsulated and had a hard consistency. Histologicalexamination of the excisedspecimenconfirmed the diagnosis of intra-orbital cavernoushemangioma. The immediate post-operative course wasuneventful, withcompleteregression of the leftexophthalmos. Palpebralecchymosis and leftchemosisappearedafter the surgery but resolvedcompletelywithin a few days. After a two-year follow-up, the patient showed no signs of local recurrence.

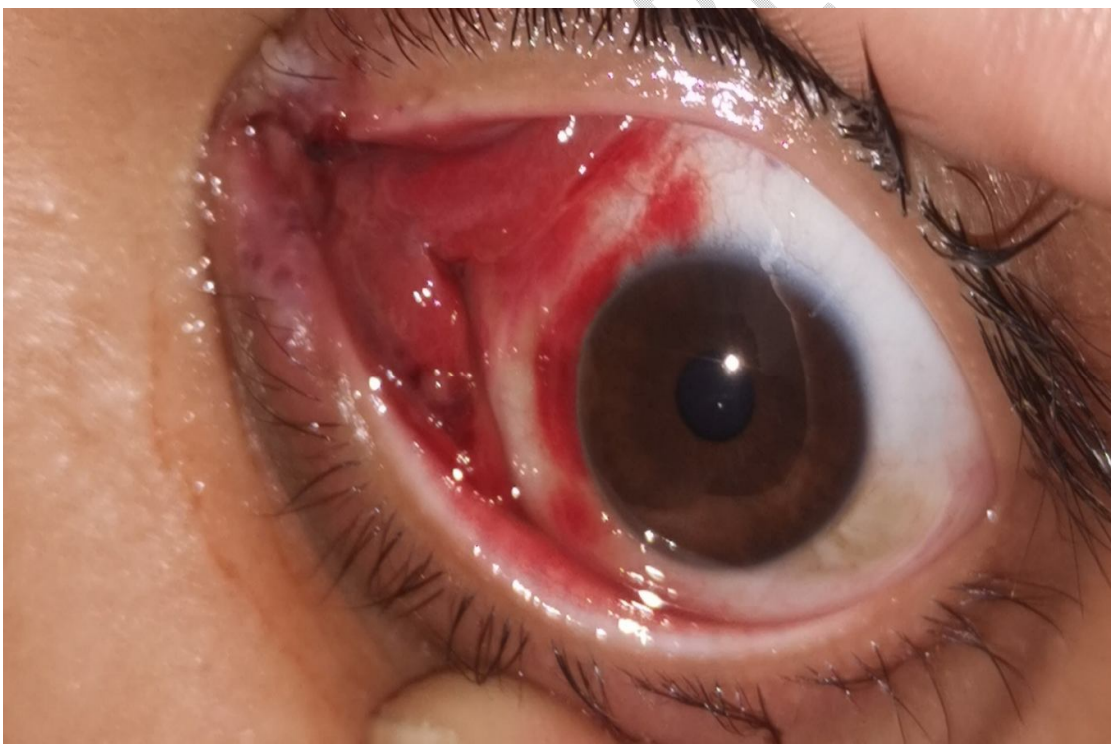

Figure 1 : A brightpink-red lesion in the medial canthus

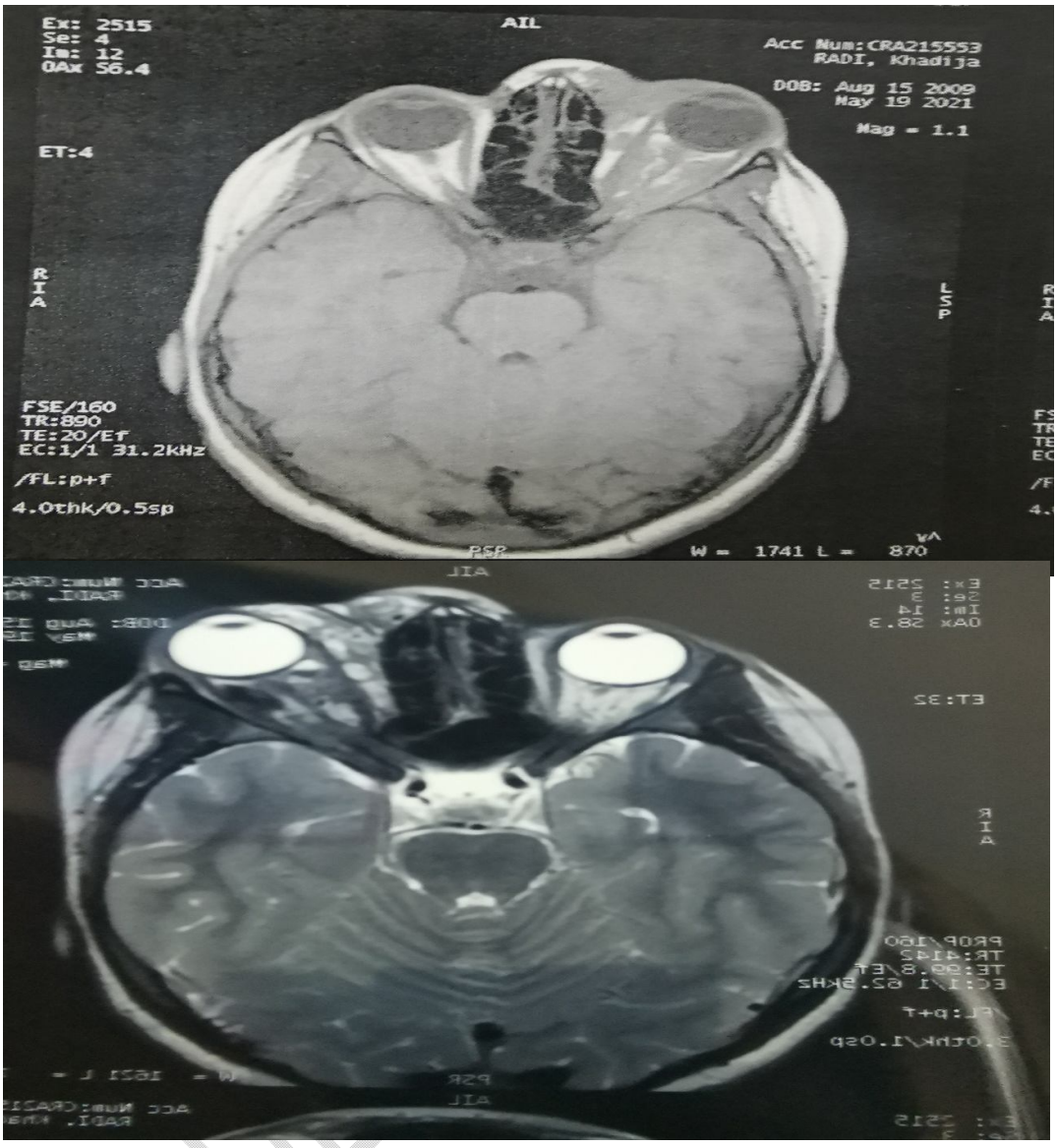

Figure 2 :MRIappearance consistent withleft intra- and extra-orbital conical hemangiomawithmedialcanthalpredominance

Discussion

Vasculartumors of the orbit are mostlybenign, and the mostcommon are cavernoushemangiomas. According to the main seriespublished [1], cavernoushemangiomasaccount for 3-14% of all orbital tumors, and 50-80% of all orbital vasculartumors [2]. Theygenerally affect youngadults, rarelybefore the age of 20. However, our patient presentedwiththistumor at an olderage, with a clearfemalepredominance. In fact, the sex ratio isseven out of tenwomen. Common symptoms of thesetumorsinclude axial exophthalmos, whichis progressive, painless, reducible, non-pulsatile and

maybeaccompanied by decreasedvisualacuity or ocularmotilitydisorders. In rare cases, a more acute presentationwithhaemorrhage or thrombosis has also been reported [3].

Thesetumors are usuallyunilateral and can vary in location and volume. The main treatmentissurgical, but the location of the lesion and itsrelationshipwithintraconicalvascular-nervous structures make the procedurecomplex.

There are three main therapeutic options for treatingthesetumors: (4) intratumoral injection of corticosteroids, whichmay use differentmoleculesjointly (triamcinolone, betamethasone, dexamethasone and methylprednisolone); (5) surgical excision; (6) oral corticosteroidtherapy (Prednisone® 1 mg/kg/day) with the usualassociatedmeasures (low-saltdiet, vitaminocalcium and potassium supplementation) [4, 7]. Thesetherapeutic options can beusedalone or in combination. In cases where a single therapyis not effective, thesedifferent options can beusedsynergistically.

REFERENCES

1. P.-A. Aymarda, ∗, B. Langloisa, M. Puttermanb, P.-V. Jacometa, S. Moraxa, O. Galatoirea, Management of orbital cavernoushemangiomas — evaluation of surgicalapproaches: about 43 cases, French Journal of Ophthalmology (2013) 36, 820—829

2. J. Bouguila 1, *, K. Yacoub 1, H. Bouguila 2, N. Ben Neji 1, S. Sahtout 1, G. Besbes 1. Intra-orbital cavernoushemangiomas. revStomatol Chir Maxillofac 2008;109:312-315

3. uban JM, Navailles B, Yalazkan B. Large orbital-palpebralcavernoushemangioma. J Fr Ophthalmol1992;15:357—62.

4. 1. Wasserman BN, Medow NB, Homa-Palladino M, Hoehn ME. Treatment of periocularcapillaryhemangiomas. J AAPOS 2004;8:175-81.

5. Offret G, Brini A, Dhermy P, Bec P. Pathologicalanatomy of the eye and its appendages. Report from the French Society of Ophthalmology. Éditions Masson; 1974.

6. Brini A, Dhermy P, Sahel J. Oncology of the eye and adnexa. Anatomical-clinical atlas. Dordrecht; Boston; London: Kluwer academicpublishers; 1990.

7. Offret G, Haye C. Tumors of the eye and ocularadnexa. Éditions Masson; 1974.