INTRAOCULAR TUMOR DIAGNOSED TWO YEARS AFTER PERFORATING TRAUMA

TUMOR INTRAOCULAR DIAGNOSTICADO DOS AÑOS DESPUÉS DE UN TRAUMA PERFORANTE

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ABSTRACT

Case report: We describe the unusual diagnosis of a ciliary body medulloepithelioma by histopathology of a subretinal membrane obtained from vitreoretinal surgery of a 10-year-old boy. The patient had a history of perforating trauma OS 2 years earlier, and both fundus exam and B-scan ultrasound revealed only a retinal detachment with a subretinal membrane. No detectable mass was present.

Discussion: The membrane removed from underneath the peripheral retina revealed a blue cell tumor confirmed by histopathology and immunohistochemistry to be a primitive neuroectodermal tumor. Currently, the patient has been followed for 5 years with no signs of recurrence (Arch Soc Esp Oftalmol 2009; 84: 469-472).

Key words: Ocular trauma, retinal detachment, vitrectomy, medulloepithelioma, pathology, retinoblastoma.

RESUMEN

Caso clínico: Se describe el caso histopatológico poco común de un medulloepitelioma del cuerpo ciliar, encontrado en una membrana sub-retiniana, que se obtuvo durante la cirugía vitreo-retiniana de un niño de 10 años de edad. Dicho paciente tenía un historial de haber tenido un trauma perforante en el ojo izquierdo dos años antes. Tanto el examen oftalmoscópico como el ultrasonido revelaban sólo un desprendimiento de retina con una membrana sub-retiniana. No había evidencia de una masa intraocular.

Discusión: La histopatología y los estudios de inmunohistoquímica de la membrana extraída de la retina periférica, revelaron un tumor de células azules, compatibles con un tumor primitivo de origen neuroectodérmico. Al momento, el paciente ha tenido cinco años de seguimiento sin recurrencia alguna.

Palabras clave: Trauma ocular, desprendimiento de retina, vitrectomía, medulloepitelioma, patología, retinoblastoma.
INTRODUCTION

The usual management of perforating trauma with secondary retinal detachment is pars plana vitrectomy with intra-ocular buffering with or without scleral indentation (1). And though modern vitreoretinal surgery techniques allow for a fixation of retinai, many factors limit the recovery of visual acuity of children in amblyopiogenic age (1). Studies with mode B echography and computerized tomography are necessary because the risks of local and systemic malign dissemination related to vitrectomy surgery in children with media opacity which subsequently confirmed intra-ocular tumors are well known (2).

In addition, the medulloepithelioma diagnostic is not always evident in an isolated clinical exploration (2-4). The most frequent symptom of medulloepithelioma is no vision followed by pain and signs such as tumor-like mass in the iris or ciliar body and leucochoria (2).

CLINICAL CASE

A 10-year-old child with a history of perforating trauma in the left eye (LE), sutured immediately after the event two years earlier. The ophthalmological follow-up was regular. Visual acuity was of 20/20 in the healthy eye and light perception in the LE. The ophthalmological exploration revealed in the LE temporal corneoscleral scar extending between six and 12 o’clock, associated to anterior and posterior synechiae (fig. 1). A partially reabsorbed cataract prevented the observation of said eye. Intra-ocular pressure was of 12 mmHg. The patient was recommended a cornea transplant combined with cataract surgery and intra-ocular lens implant for recovering vision.

A pre-op mode B echography facilitated the identification of retinal detachment associated to subretinal membrane and traction in the vitreous (fig. 1). Immersion technique exploration did not reveal ocular or arbitrary alterations. Orbits and brain OCT were normal. It was suggested to extract a cataract associated to posterior vitrectomy, temporal retinotomy, excision of subretinal membrane, perfluorocarbon injection, endophotocoagulation and fluid-oil exchange.

The small membrane fragments with bronze-colored pigment dispersion were obtained under the retina. The largest fragment measured 7x2x1 mm. The microscopic exploration of the fragments revealed a fibrous tissue with pigmented cell groups compatible with retina pigmented epithelium. Adhered to the edge of the membrane we observed an area of blue cells with neoplastic characteristics which included an alteration in the nucleus-cytoplasm relationship, nuclear modeling, cellular pleomorphism, pseudo-rosettes and mitosis (fig. 2). The different branch of diagnostic included retinoblastoma and medulloepithelioma. Four immuno-histochemical stains were made: protein S-100, neuronal specific enolase (NSE), glyal fibrillary acid protein (GFAP) and synaptophysine (fig. 2).

The patient remained without symptoms for 40 days, after which period the intraocular silicone emulsified. The patient was then submitted to additional surgery to change the oil and remove the remains of the tumor. All the obtained material was negative for blue cell tumor. Even though the natural history evolution of the eye made it become phthysic, the parents did not authorize enucleation. The patient remained under ophthalmological and oncological observation for a five-year period after the second vitrectomy without exhibiting local or systemic signs of tumor relapse.

DISCUSSION

Medulloepitheliomae are rare congenital embryonic tumors reported in children and adults (3,5). The clinical diagnostic is not always apparent. The echographic localization is usually difficult because these tumors exhibit flat and expanded growth, which could explain the post-op diagnostics (2,4).

The histopathological characteristics of intraocular medulloepitheliomae are varied (3-5). These tumors can be classified as teratoid and non-teratoid medulloepitheliomae. The main characteristic of the non-teratoid type is the multiple layers and strings of poorly differentiated neuro-epithelial cells simulating an embryonary retina or ciliar epithelium (3).

The differential diagnostic includes retinoblastoma, particularly when the posterior vitrectomy has been performed because the increased possibility of dissemination and arbitrary invasion is well known (2). Even though medulloepitheliomae can exhibit large neuroblastic pseudo-rosettes, these consist in several tubular-structure cell rows with-
out exhibiting the handle or glove-shaped growth pattern of retinoblastomas. They can be differentiated from the Flexner-Wintersteiner rosettes due to the relationship with cellular union and the absence of pseudo-photoreceptors (3).

The immunohistochemistry of medulloepithelioma with pseudo Flexner-Wintersteiner rosettes is always positive for NSE and synaptophysine as in this case (3). Although approximately 40% of medulloepitheliomae are malign, our patient exhibited several characteristics suggesting a benign nature, confirmed by five years of follow up without relapses.

Another medulloepithelioma diagnosed by histopathology in a perforated eye has been published (5). Said eye was enucleated due to infection and conjunctival and orbital extension, which did not occur in our patient.

It is important to bear in mind that neoplastic processes without pigmentation in the ciliar epithe-

lum are divided in embryonal and non-embryonal. Embryonal tumors are medulloepithe-

liomae with different presentations as discussed above. Non-embryonal tumors are adenomas/ ade-

nocarcinomas, reactive and pseudo-adenomatous hyperplasia. As a result of the trauma it can be stated that ciliar epithelium reactive hyperplasia was the most probable finding. However, histopathological findings excluded said possibility.

Finally, this case became intriguing because we tried to conjecture on a possible relationship...
between the perforating trauma and histopathological findings two years later. Considering these facts, at least three questions must be pondered: Did the left eye have sufficiently low vision beforehand to facilitate the trauma? Did the trauma trigger the appearance or proliferation of neoplastic cells or were these already present? Assuming the preexistence of neoplasia, why didn’t the perforation promote the dissemination of the tumor? These questions have yet to be answered. However, the unusual characteristic of this case emphasizes the importance of histopathological analysis of all surgically removed ocular tissue to guarantee the patient the best performance and quality of medical practice.

REFERENCES


Fig. 2: Histopathological findings: Microphotograph of removed sub-retinal membrane showing a fibrous chain adhered to and islet of blue cells at one end (upper right). Large magnification microphotograph of neoplastic cells (upper left). Below, the images show a positive stain for neuronal specific enolase (NSE) and synaptophysine (the magnifications are indicated in each photograph).