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Adaptation of a Hydrogel Expander and Rigid Conformer: The Case Report of a Girl with Retinoblastoma

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Abstract: Ocular conformers are considered the first stage of treatment to enlarge the cavity and these can be static or dynamic. The expander that we propose are hydrogel balls that in their natural state have a size of 3 mm. and they are osmotic filled, becoming hydrated up to 18 mm with tears and moisture from the patient's orbital cavity. The objective is to use an osmotic hydrogel former and demonstrate its evolution phase during treatment. The methodology will be of an exploratory and experimental descriptive type where the hydrogel former will be applied to the patient and the evidenced results will be described step by step, in the experimental part the rehabilitation effects of the orbital cavity will be evaluated. As a result, it is reported that by adapting the osmotic hydrogel conformer, it was possible to expand the ocular cavity and insert a provisional ocular prosthesis. It is concluded that using an osmotic-type hydrogel former, it was possible to expand the girl's cul-de-sac, which shows a great therapeutic help in the process of adapting personalized ocular prostheses.

Keywords: Retinoblastoma, Anophthalmia, Conformer, Hydrogel, Osmotic

1. Introduction

The osmotic-type hydrogel expanders that have been created by the OC. Sergio Osan from the city of Mendoza, Argentina, contribute enormously within the professional practice in the adaptation of ocular prostheses for all types of patients who present the anophthalmic condition for any reason, even more so if they are infants, since treatment can be started from of the fourth month of birth. These devices are easy to use since it involves inserting a ball into the cavity so that it is embedded with the patient's own tear, guaranteeing an orbital pressure of no more than 30 mm/hg. [1] What is going to fill the spaces of the ocular cavity gently without causing internal damage (lacerations). The Expanders that we are proposing in this study are balls of pure hydrogel [2] that in their natural state without hydration have a size of 1 to 3 mm. They are osmotic filled, that is, they are hydrated with the same tear and the humidity of the patient's orbital cavity and are adapted in the same offices, they do not need sutures.

Figure 1. *CEPROC hydrogel ball in osmosis process, photography by Efraín Silva 2021.*

Retinoblastoma

Retinoblastoma is a cancer in the retina of the eye. The retina is the light-sensitive lining at the back of the eye. It can affect one or both eyes. It causes a tumor to form inside the eye, and can grow and damage internal structures. [3, 4]

Causes of Retinoblastoma

Retinoblastoma is a rare childhood cancer that affects about 300 children in the United States each year. Although retinoblastoma can be diagnosed at any age, most children are diagnosed before the age of 2 years, and most cases are

diagnosed in children younger than 5 years. Retinoblastoma appears to affect boys and girls equally, as well as African-Americans and Caucasians. Children who have a parent or sibling with this disease or children with a known chromosome 13q mutation are at increased risk of developing retinoblastoma.

Retinoblastoma occurs when there is a mutation in the retinoblastoma gene (RB or RB1). This is a tumor suppressor gene that acts as a brake on cell division. This gene is present in all cells of the body. There are two copies of the RB1 gene in each cell. This gene is located on chromosome 13q. For a retinoblastoma to occur, both copies of the gene must have the mutation. [3, 5]

The cause of this cancer is a change in a gene. It may be a gene passed down from parents (inherited). Or it may be a genetic change that occurs randomly (sporadic).

In 1 out of 3 children with retinoblastoma, it is present at birth (congenital). Of these children, 1 in 4 has a parentally transmitted form. It usually affects both eyes. It also increases the risk of other types of cancer, such as sarcoma and melanoma.

In 2 out of 3 children with retinoblastoma, the disease occurs randomly. In these cases, it only affects one eye. There is no increased chance of having other types of cancer [5].

Signs and Symptoms

Patients usually present with leukocoria (a white reflection in the pupil, sometimes called a cat's eye pupil) or strabismus. Much less often, they have inflammation of the eye or impaired vision.

Rarely, the cancer has already spread through the optic nerve or choroid or via the bloodstream, causing an orbital or soft tissue mass, local bone pain, headache, anorexia, or vomiting.

When the diagnosis is suspected, the fundus of both eyes should be thoroughly explored by indirect ophthalmoscopy, with the pupil dilated and the child under general anesthesia. Cancers are manifested by isolated or multiple gray-white elevations in the retina; tumor seeding can be seen in the vitreous. [4, 5]

Retinoblastoma is diagnosed by direct observation of tumors within the eye, ultrasound, RetCam imaging, and magnetic resonance imaging. [6]

International Classification

Group A

Small tumors (less than 3 mm) that are only on the retina and more than 3 mm away from the foveola (the center of the fovea) and more than 1.5 mm away from the optic disc.

Group B

1) Tumors greater than 3 mm that are limited to the retina in any location.
2) Clear subretinal fluid less than 6mm from the edge of the tumor.

Group C

1) Localized vitreous and/or subretinal spread (less than 6 mm from the tumor margin).
2) There are no tumor masses, lumps, or snowballs in the vitreous or subretinal space.

Group D

1) Diffuse vitreous and/or subretinal spread (more than 6 mm from the tumor).
2) Subretinal fluid more than 6 mm from the tumor margin.

Group E

No visual potential or presence of one or more ofthe following:

1) Anterior segment tumor.
2) Tumor in or on the ciliary body.
3) Neovascular glaucoma.
4) Vitreous hemorrhage obscuring the tumor or hyphema important.
5) Consumptive or pre-consumptive eye.
6) Presentation of orbital cellulitis type [6].

Types of Retinoblastoma

Congenital (Hereditary)

1) A child with congenital retinoblastoma has an abnormality in the RB1 gene. Congenital retinoblastoma occurs in two ways. This mutation can be inherited from a parent, even if they have never had a retinoblastoma: they are "carriers" of the mutated gene, or there is a germline mutation of these genes during fetal development. A germline mutation is any change in the cells that are developing after conception.
2) Children with congenital retinoblastoma are at increased risk of developing retinoblastoma in both eyes and may also have multiple tumor sites within the eye.
3) They may also have an increased risk of other types of cancer, such as pineoblastoma, a tumor of the pineal gland at the base of the brain.
4) Occurs more frequently in young children.

Sporadic (Not Hereditary)

1) It is also produced as a result of the RB1 gene, but only in one cell of the eye.
2) Typically, these children only develop a tumor in one eye.
3) May occur more frequently in older children. [3].

Treatment

1) For unilateral cancer, enucleation.
2) For bilateral cancer, options are photocoagulation, intra-arterial chemotherapy, or unilateral enucleation with photocoagulation, cryotherapy, and irradiation of the other eye.
3) systemic chemotherapy.

It is necessary to repeat the ophthalmological examination of both eyes and, if necessary, the treatment, with intervals of 2 to 4 months. [4, 3]

Presentation of the case

CLINICAL DATA: Patient with decreased visual acuity of the right eye.

SAMPLE ORIGIN: Right eyeball

DATE: 04/05/2021

Macroscopic Description

Right eyeball measuring 2.3 x 2 x 2 cm, which at the posterior level shows an ocular nerve of 0.7 cm, which on cut shows a tumor lesion at the level of the posterior chamber of 1.5 cm with a whitish appearance, apparently no ocular wall infiltration.

Microscopic Description

Histological sections show eyeball where malignant neoplastic tissue is observed at the level of the posterior chamber, characterized by the proliferation of round blue cells, with hyperchromatic nuclei, slightly pleomorphic with little cytoplasm arranged diffusely, with formation of images in pseudo rosettes around of capillaries, with invasion of the macula.

The ciliary body and muscle are free of tumor cells, without invasion of the anterior chamber.

The choroid, sclera, lens are free of tumor infiltrate. The foveola, the optic nerve with the surgical border, soft tissues, muscle and peri-ocular adipose tissue are free of tumor infiltration.

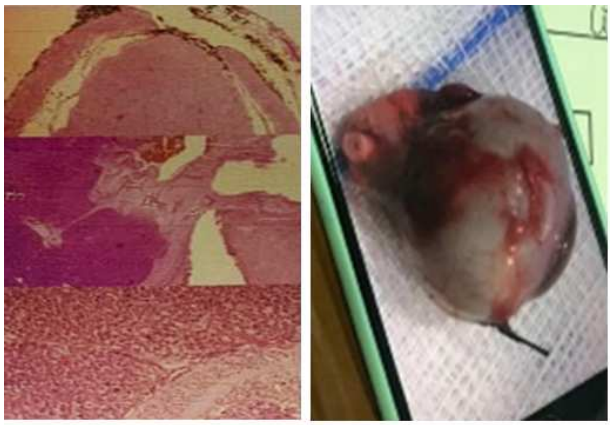

Figure 2. *a) Microscopic image of the patient. b) Image of enucleated right eye. Image source Dr. Francisco de Icaza Bustamante Children's Hospital 2021.*

Histopathological Diagnosis

Right eyeball resection product:

1) RETINOBLASTOMA, located in the posterior chamber, with involvement of the macula.
2) ANTERIOR CHAMBER, body and ciliary muscle free of tumoral lesion.
3) CHOROID, sclera and lens are free of tumor.
4) FOVEOLA, optic nerve and periocular soft tissues free of tumor.

CLINICAL DATA: Patient with diagnosis of retinoblastoma

SAMPLE ORIGIN: Bilateral bone marrow

DATE: 20/05/2021

Macroscopic Description

LABELED 1 Fragment of elongated cylindrical tissue measuring 0.8 cm brownish yellow of firm consistency.

LABELED 2 Fragment of elongated cylindrical tissue measuring 0.7 cm brownish yellow with a firm consistency.

Microscopic Description

1 and 2 the histological sections show fragments of bone spicules and hyaline cartilage whose hematopoietic content presents the three series, with a good cellular and adipose relationship with normal histological characteristics according to the patient's age.

No malignant neoplastic changes are found in the examined sample.

Histopathological Diagnosis

1 and 2 right and left iliac crest puncture biopsies: Normocellular bone marrows free of neoplastic infiltration.

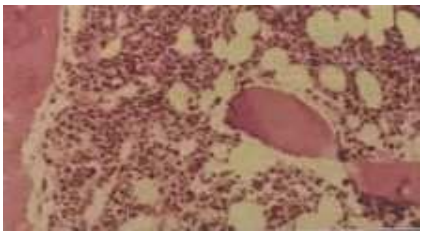

Figure 3. *Microscopic image of the patient. Image source Dr. Francisco de Icaza Bustamante Children's Hospital 2021.*

Once the right eye has been enucleated in order to preserve the aesthetic part and avoid sagging of the cheekbones and loss of orbital fat, the 1-year-3-month-old female patient goes to the Optical Silva de la Ciudad de Guayaquil for consultation accompanied by her mother in search of an ocular prosthesis. All the corresponding tests are carried out, starting with the clinical history and informed consent, in order to start with the physical examination. In the case of the right eye, a visualization technique is performed to observe the internal structures and see the best possible treatment. that he had one month after having enucleated his eye.

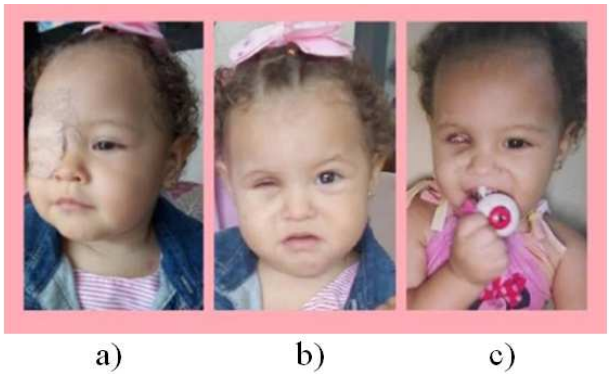

Figure 4. *A one-year-old, three-month-old patient with enucleation of the right eye due to retinoblastoma can be seen, a) the patient's condition when she arrived b) how she was left a month after surgery c) the patient with the CEPROC hydrogel ball). Image source Efraín Silva 2021.*

Analyzing the case of the girl whose clinical history reveals enucleation of the right eye due to retinoblastoma. The respective indications are given to the Mother of the Patient, to whom the use of the CEPROC osmotic ball is suggested, since at the time of visualizing the internal structures, the folds and the bottom of the sac are observed to be stuck together. This is the reason why this painless treatment is chosen, without the risk of lacerations for eight continuous hours of use. After the suggested time has elapsed, it can be verified that the internal structure of the orbital cavity was fully enlarged, which facilitated the insertion of a rigid former, for a period of fifteen days for the placement of the personalized prosthesis.

Said prosthesis must be controlled periodically and after approximately two years it must be changed due to the anatomical changes that the patient will present in her orbital and facial parts.

a) b)

Figure 5. *a) image of the patient with the initial rigid former for 20 days. b) image with the personalized prosthesis in polymethylmethacrylate material and plasma treatment. Photography by Efrain Silva 2021.*

2. Discussion

The procedure that was carried out with this patient is based on retrospective studies verified in Argentina with the OC. Sergio Osan scientifically [8, 5] the benefits of treatment with osmotic-type conformers [9, 10] are innumerable, especially from our point of view, it is the early start of treatment without causing trauma in infants, the enlargement of the cul-de-sac in the orbital cavity is almost immediate without causing any pain since the pressure of the ball is very low, the benefit of using the ball is that it does not need to be changed constantly since the ball is imbibed from the patient's own tear which helps in asepsis and is also harmless. For the treatment of this type of patients, it must be carried out with the help of a multidisciplinary team, a psychologist for their rehabilitation in the emotional part and self-esteem, the Ophthalmologist to treat possible infections that could occur due to the use of prostheses without due care. and the Optometrist in the Adaptation, readjustment, cleaning and maintenance of the prosthesis [11]. What is significant about this case is the presence of retinoblastoma, being a disease that could even lead to the death of the patient if it is not operated on time. The advantage of treating it on time is the total removal of the cancer and subsequently treating the cavity with the same prostheses that in early stages help in aesthetic recovery and good results are achieved by carrying out strict controls within the treatment in the expansion phase of the cavity. and depth of the cul-de-sac. In the case of the patient, it was possible to adapt the personalized prosthesis in just two months of treatment.

3. Conclusions

This is a one-year-three-month-old girl who was diagnosed with cancer of the right eye (retinoblastoma), which was removed in its entirety and then began with the adaptation treatment of a personalized ocular prosthesis, which had several phases beginning by the adaptation of an osmotic-type hydrogel ball for eight hours of uninterrupted use, thereby achieving the enlargement of the cul-de-sac and the stretching of the conjunctival folds, since after surgery it was occluded by means of a patch and there was no type of stimulation. The next phase was adapted to a rigid polymethylmethacrylate [11] former with eye drawing and ocular lubricant for twenty days for its subsequent evaluation, which was successful and once the final mold was stabilized, the personalized prosthesis was elaborated with plasma treatment [12] to avoid possible adherence of microorganisms in the porosities of the rigid material, which guarantees better asepsis and easy handling by the user, we believe that the hydrogel former expands with pressure in a uniform manner and fills all the spaces empty at the orbital level, in addition to helping the conjunctival folds to relax to achieve a better cul-de-sac and to be able to adapt the ocular prosthesis, it should be noted that the hydrogel former is harmless, that is, it is not toxic since it is soaked with the same tear of the patient in addition to keeping it always moist, which facilitates better blinking, avoiding friction or lacerations with mo those that are common with rigid formers.

Conflict of Interest

The authors declare that they have no conflict of interest.

Acknowledgements

The authors extend their thanks to the girl's parents for allowing and wishing that this case of retinoblastoma be disclosed and the initial treatment with an osmotic-type conformer and finally the use of a prosthesis delivering all the clinical history reports, in addition to allowing the publication of the images, to the specialized department of adaptation of ocular prostheses of the Óptica Silva of the City of Guayaquil, Ecuador [13] and to the Center for Ocular Prostheses and Contactology Specialized (CEPROC) [14].

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