Case Report

A Prosthetic Approach for Rehabilitation of a Pediatric Patient with Retinoblastoma

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Abstract

Retinoblastoma is a highly malignant tumor of an eye, which is often detected at a far advanced stage in childhood. The most preferred treatment for such neoplasm is enucleation followed by placement of orbital implants. The affected child undergoes overwhelming psychological, social, emotional, and behavioral changes due to facial asymmetry resulting from enucleation. A team approach including pediatric ophthalmologist, psychologist, and a maxillofacial prosthodontist plays an important role in such cases helping such children in their reintegration in the society. This paper presents a case of a 4-year-old male patient who had undergone enucleation of his right eye due to intraocular retinoblastoma. The patient was rehabilitated cosmetically with precisely fabricated customized ocular prosthesis. An ocular prosthesis is a highly positive and noninvasive approach to improve the cosmetic appearance and psychological well-being of the patient.

Keywords: Custom ocular prosthesis, enucleation, pediatric, retinoblastoma

Introduction

"Cancer" is always a frightening and life-threatening diagnosis, and when it affects children, the situation is overwhelming. Eye cancer in children, though rare, is the third leading cancer of childhood. Retinoblastoma, a cancer of the retinal eye tissue, is a highly malignant tumor of the eye that is potentially fatal and can result in vision loss of one or both eyes.[1] It affects almost all races with equal sex predilection with an incidence varying from 1:10,000 in South Africa to 1:34,000 in the Netherlands.[2,3] This malignancy accounts for approximately 2.5%–4% of all cancers diagnosed in children younger than 15 years.[4] Retinoblastoma is of two types, hereditary retinoblastoma (familial) which is inherited from parent to child and is bilateral and the other form is nonhereditary retinoblastoma (sporadic) which is unilateral carrying no increased risk of a second tumor. The hereditary form of retinoblastoma, responsible for 30%–40% of the cases, is associated with a long-term predisposition to other types of cancer.[5] About 90% of all retinoblastoma cases are diagnosed within the first 3 years of the child's life, whereas, children with familial retinoblastoma typically are diagnosed at an earlier, i.e., 4 months of age. When there is no family history, the retinoblastoma is usually not diagnosed by both the family and the physician. The most common and early clinical sign (56%) seen in an eye with retinoblastoma is the presence of a white papillary reflex (cat's eye reflex) or leukocoria.[6] This sign may be noticed by a parent after a flash photograph is taken and also might be noted by the child's physician during a routine eye checkup. The other sign is lazy eye (strabismus) in which the eyes do not appear to look in the same direction. This may be caused by a mild weakness of the muscles that control the eyes, but it can also be caused by retinoblastoma. Numerous methods employed routinely for diagnosing retinoblastoma include transillumination, fluorescein angiography, ultrasound (echography), optical coherence tomography, computed tomography scan, magnetic resonance imaging, biopsy, lumbar puncture (spinal tap), and bone marrow aspiration.[7]

Early diagnosis and immediate treatment are crucial parameters in saving child's life and in salvaging useful vision in one or both eyes. However, unilateral retinoblastoma is usually far more advanced at the time of diagnosis, and bilateral

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retinoblastoma is often asymmetric in presentation, with one eye involved by massive tumor and the other one less involved.[7] Retinoblastomas can be staged into two main groups: intraocular retinoblastoma, in which the cancer is still within the eye, and extraocular retinoblastoma, in which the cancer has spread beyond the eye. Extraocular cancers can be divided further into orbital retinoblastomas, which have spread only to the eye socket, and metastatic retinoblastomas, which have spread to distant parts of the body, such as the brain or bone marrow. Different approaches to treat intraocular retinoblastoma include enucleation, laser photocoagulation, laser hyperthermia, cryotherapy, radioactive plaque brachytherapy, external beam radiotherapy, systemic chemotherapy, and stem cell transplant.[8] Sometimes, more than one type of treatment may be used. The treatment approach is dependent on the size and extent of tumors, the involved sites, and child's systemic involvement. Enucleation is the most preferred treatment for retinoblastoma in which the whole eye and the optic nerve attached to it are surgically removed.[9,10] During the same operation, an orbital implant (made of silicone or hydroxyapatite) is usually placed to take the place of the eyeball. It is attached to the muscles that moved the eye, so it should move the same way as the eye would have. The most obvious drawback of enucleation is the loss of vision and hindrance of future growth of bone and other tissues around the eye socket. Hence, ocular prosthesis should be fabricated as early in the childhood life as possible.[11] Enucleation can make the area look somewhat sunken that can be prevented by fabricating an ocular prosthesis of a larger size from time to time as child grows.[12] The psychological effect of loss of an eye may pose great problems for the child than the physical disability.[13] Apart from loss of vision, the child also faces a distortion in his facial appearance. Such child requires an immediate attention and utmost care from specialists during rehabilitation. A multidisciplinary approach is most appropriate to rehabilitate such child in terms of psychological, social, and emotional aspects. A team of specialist including a pediatric ophthalmologist, plastic surgeon, psychologist, and maxillofacial prosthodontist are essential in restoring the child's quality of life.[14] This case report presents a prosthetic approach to rehabilitate a 4-year-old boy with retinoblastoma, who underwent enucleation of his right eye.

Case Report

A 4-year-old male patient reported to the department of prosthodontics with the complaint of unesthetic facial appearance due to the absence of his right eye [Figure 1]. His parents gave history of retinoblastoma which was diagnosed at the age of 2 years, for which, enucleation was performed at 2.5 years of age. Patient also received chemotherapy and radiotherapy for treatment of his cancer. At the age of 3 years, a stock artificial eye was placed in the patient's eye; but, due to pain and irritation caused by artificial eye, patient used to remove the prosthesis and showed extreme resistance to wearing of the prosthesis. Patient's previous experience with prosthesis revealed his disliking toward the wearing of stock ocular prosthesis. After careful examination of eye socket, it was planned to fabricate a custom ocular prosthesis for the patient. All the details of procedure, including its maintenance and limitations, were discussed to the patient and his parents to gain their cooperation. A written consent was obtained for making photographic records.

Clinical procedure

The treatment planning was initiated by making the impression of the eye socket with the help of a thin mix of ophthalmic alginate impression material (Ophthalmic Moldite, Milton Roy Company, Sarasota, FL, USA). This impression was poured in dental plaster, and a primary cast was obtained. A tray (DPI cold cure; Dental Products of India, Mumbai, India) was fabricated on this cast to make a definitive impression. The patient was instructed to gaze straight ahead while making the impression. The light-viscosity elastomeric impression material (Reprosil; Dentsply DeTrey GmbH, Konstanz, Germany) was slowly injected into the socket taking care to avoid any air bubbles [Figure 2]. The patient was instructed to make various eye movements to make functional impression of the eye. The impression material was reinforced with a syringe needle cover to hold it in place and for ease of removal after it sets. The impression was then poured with dental stone (Kalabhai Karson Pvt. Ltd., Mumbai, India) to obtain two-piece cast by using double-pour technique [Figure 3]. The impression was separated from the cast and lubricated the stone cast with a thin coating of vaseline. The socket of the cast was filled with molten wax, and after solidification, the retrieved wax form was flasked and processed in tooth-colored acrylic (Trevalon, Dentsply India Pvt. Ltd.) prematched with natural sclera of the contralateral eye. For matching sclera shade, the shade tabs were prepared by mixing and matching different shades and proportions of tooth-colored acrylic till the color of sclera of the other eye was replicated. The processed sclera blank was retrieved from the flask so that the stone matrix is preserved for final processing of the globe's original contour. The highly polished sclera blank was placed in the patient's socket for evaluation, and necessary adjustment was done until the normal corneal contour is simulated. The patient was instructed to hold an erect position and to gaze straight ahead and observed from the side to decide the iris plane

Figure 1: Preoperative photograph showing enucleated socket of the right eye.

relationship with the normal eye. The distance from the pupil of the normal eye to the midline was used in determining the horizontal position of the prosthetic pupil center and marked on the globe. Similarly, the vertical position of the pupil center was established and marked by the canthus relationships. This way, the papillary center was established and marked on the globe and was then removed. The diameter of iris, as measured from the normal contralateral eye, was circumscribed on the globe with a compass [Figure 4]. Return the globe to the socket and the outlined iris evaluated in relation to that of eyelids. Accurate iris positioning is critical in establishing natural appearance. The globe was then modified by cutting away resin within circumscribed area to provide a chamber to house the selected iris from the stock shell. The selected iris was leuted with clear autopolymerizing resin, and minor characterization was done. Final evaluation was done in the patient, and the characterized globe form was returned to its original position within the stone matrix of the flask. The remaining space was packed with clear heat-polymerizing resin (Trevalon, Dentsply India Pvt. Ltd.) and was processed in the conventional manner. After processing, the properly finished and polished prosthesis [Figure 5] was inserted in the socket after being disinfected with 0.5% chlorhexidine and 70% isopropyl alcohol for 5 min and lubricated with an ophthalmic lubricant (Ecotears, Intas Pharmaceuticals Limited, Ahmedabad, India) to maintain a tear film over the prosthesis and to improve eye movements. Minor adjustments were made at the time of delivery as per the patient's comfort and esthetics [Figure 6]. Instructions were given to the patient regarding proper care and handling of the prosthesis as well as the maintenance of socket hygiene, and the need for regular recall appointments was emphasized. As in the early childhood, orbital volume increases in a linear fashion[15,16] and a change of prosthesis is required between 18 and 26 months following prosthesis placement in children.[17] Hence, the patient was recalled every 6 months for follow-up. In the follow-up period of 6 months, the patient was able to wear a cosmetically acceptable ocular prosthesis without any discomfort. Hence, the patient was successfully rehabilitated by custom ocular prosthesis.

Figure 2: Impression of the socket with light-viscosity elastomeric impression material.

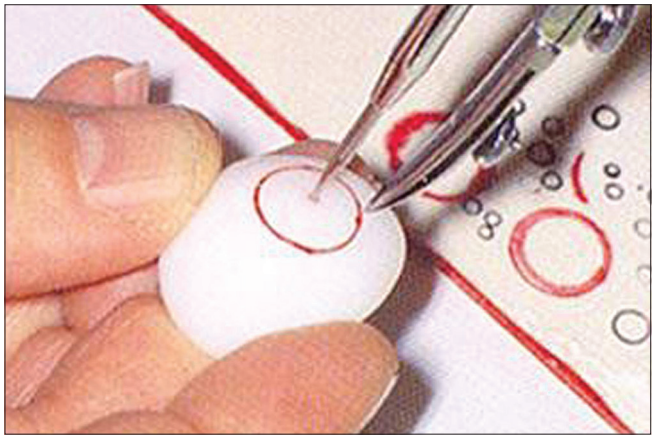

Figure 4: Proper papillary dimensions circumscribed with a compass.

Discussion

Any type of retinoblastoma can prove a fatal form of eye cancer, if undetected, and can lead to loss of vision due to loss of one or both eyes. This loss often results in an overwhelming number of psychological, social, and emotional responses in the affected patient. Enucleation during childhood or congenital anophthalmos or severe microphthalmos often causes hindrance in normal bony orbital growth, facial

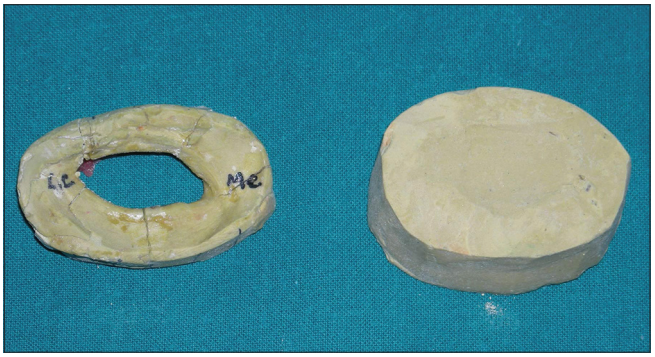

Figure 3: Two-piece working cast obtained with double-pour technique.

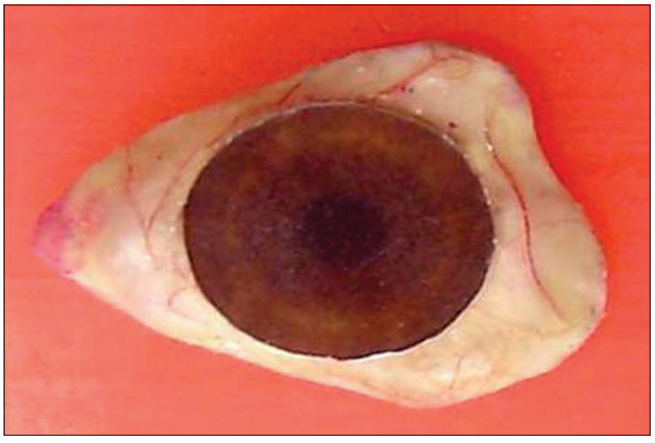

Figure 5: Color characterization and globe completion.

Figure 6: Postoperative photograph showing prosthesis in lieu of enucleated eye.

distortion, and asymmetry.[18] Immediate attention and treatment planning plays an important role in such patients. Customized ocular prosthesis has shown to play a definite role in stimulating the orbital growth. The periodic and regular replacement or modification of prosthesis is crucial to imitate a continuous stimulus for socket growth.[19] The prosthesis should reproduce the color, form, and orientation of iris so well that it would be easily acceptable by the patient in terms of cosmetic appearance. Among different techniques documented for the fabrication of ocular prosthesis, modification of stock eye gives definite benefits.[20]

Stock eyes are available in standard sizes, shapes, and colors and require no special skills or materials for fabrication of ocular prosthesis. The other advantages include its inexpensiveness and time-saving factor. However, the major disadvantages of stock eyes are its poor fit, poor esthetics, and poor eye movements. In most of the cases of enucleation, custom ocular prosthesis has proved to be beneficial in relation to its improved adaptation to underlying tissues, increased mobility of the prosthesis, and excellent esthetics due to better match of the size and color of the iris and sclera.[21] The close adaptation of prosthesis into the socket provides maximum comfort and restores full physiologic function to the accessory organs of the eye. The extra effort and time involved into fabrication of custom ocular prosthesis are advantageous for those who cannot afford other alternatives such as orbital implants and ensures a better drape of lid tissues and provides a superior natural appearance to both the patient and observer.[22] However, it involves complex painting procedures in various stages of fabrication which are quite difficult and are dependent on skills of the operator. In addition, the voids collect mucus and debris that further cause irritation to the mucosa acting as a potential source of infection.[12]

The procedure of ocular prosthesis fabrication that is explained in this case report follows the transformation of prefabricated eye prosthesis to a custom one achieving a good fit and esthetics. This benefitted in overcoming the drawbacks of prefabricated and custom-made eye prosthesis. The basic aim of this case report is to show an easy and simple technique for the fabrication of ocular prosthesis which does not depend much on artistic ability of the operator. It is relatively easy and convenient for a dental clinician to utilize this technique compared to others that consume a lot of laboratory time. The encountered limitation while performing this technique is that the operator has to depend on the availability of properly matching iris and papillary part in the prefabricated eyes. Furthermore, the long-term color stability of heat-polymerizing acrylic resin and its bonding with the stock eye will have to be monitored.

Conclusion

Loss of one or both the eyes due to retinoblastoma can be felt throughout the life of patient involving his/her family members. Eye prosthesis minimizes the possible discrepancy between the compromised and normal sides, thus contributing to balance and harmony of the facial development. Providing cosmetically pleasing custom ocular prosthesis to pediatric patient with retinoblastoma during childhood adds an inestimable psychosocial contribution to the physical benefit in overall rehabilitation. Custom-made eye prosthesis offers numerous benefits that include better mobility, even distribution of pressure, reduction in the incidence of ulceration, improved fit and comfort, acceptable esthetics, and improved facial contours. It has a good control over customizing the size of the iris, pupil, and color of the iris and sclera. It also stimulates bony orbital growth that provides long-term benefits of restoring facial symmetry and esthetics.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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