Case Report

Bilateral Primary Anophthamos: A Report of 2 Cases

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Abstract

Primary anophthalmos is a rare condition, characterized by complete absence of ocular tissues within the orbit. In this report two cases of bilateral anophthalmos seen within four weeks interval at the Jos University Teaching Hospital are discussed.

Keywords: Primary anophthalmos, facial defects, prostheses.

Introduction

Primary anophthalmos was first described 400 years ago as the absence of the globe and ocular tissues within the orbit1. It is a rare condition occurring in 0.3-0.6 per 100,000 births[1]. Patients with bilateral anophthalmos are usually associated with absence of optic chiasm, diminished size of the posterior optic path-ways, and agenesis or dysgenesis of the corpus callosum[2]. Proper growth of the orbital region is dependent on the presence of the globe which stimulates the growth of the orbit and proper formation of the lids and ocular forces[3]. The management of anophthalmos is difficult and unrewarding, but the use of cosmetic scleral shells in all cases of micro'phthalmos is useful and effective[4].

We present here two cases of primary anophhalmos seen at the Jos University Teaching Hospital over a period of four weeks. The attitude of parents to cosmetic repair is discussed.

Case Reports

Case I

A 3 weeks old female child was brought to the clinic with complaints of inability to open both eyes since birth. Attempts by the parents to open the eyes forcefully failed and so they decided to come to the hospital. The baby is a product of a full term supervised pregnancy delivered at home, after an uneventful antenatal period, labour and delivery.

Slight discharge was noticed from both eyes three days after delivery. As at the time of presentation, the child had not received any immunization. Parents are cattle rearers. Child was the 5th of the mother and there was no similar condition in the family. Parents were first cousins.

A general examination did not reveal any associated congenital abnormality. Ocular examination under anaesthesia showed normal lids with mild purulent discharge. The fornices were wide but the eyeball, muscle or fat were absent (see fig. I). Orbital B-mode ultrasound revealed no ocular tissues in both sockets. Patient was placed on chloramphenicol eye ointment to treat the discharge. The need for prosthesis in both sockets, to prevent orbital malformation was explained to both parents, but they declined further management.

Case II

A 9 month old boy was brought to the clinic by both parents with complaints of inability to see since birth. The mother noticed that the eyelids in both eyes were “glued” together, and when she separated them she discovered that, the eyeballs were absent. Parents did not seek medical attention as they resigned their fate to God, until someone advised them to come to the hospital.

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Fig. 1: Congenital anophtalmous in a 4-month old boy.



Fig. 2: Absence of eye-ball in the socket in the same patient.

The baby was a product of a full term pregnancy. Mother had fever and was treated in a Cottage hospital for malaria at 8 months of gestation. Labour and delivery were uneventful in a hospital. His milestones were appropriate for his age. He has taken all the childhood immunizations except measles. The child is the 2nd born of the parents. General examination revealed a healthy looking child. No neurological deficit was detected.

Ocular examination showed closed eyelids, narrow palpebral fissures and bilateral absence of eyeballs, muscles and orbital fats [fig. II]. B-mode orbital ultrasound revealed some undifferentiated tissues in both orbits. Parents were advised on the need to extend the palpebral fissures with the aim of inserting prostheses but they declined since there was no hope for vision for the child. They were however counseled on the need to send the child to the school for the blind when he is of school age. They accepted to send the child to the school for the blind when he is of school age.

Discussion

Anophthalmia occurs when the neuroectoderm of the primary optic vesicles fail to develop properly from the anterior neural plate of the neural tube during embryological development[1].

A study of 113 children with micropthalmia and anophthalmos showed no prenatal infections such as rubella or cytomegalovirus infection even in the 4 children with bilateral involvements[4]. In the present report, there were no suggestive cases of rubella or cytomegalovirus infections in the mothers during pregnancy. Parents of the first case were first cousins. A similar finding of consanguinous marriage has been reported in an infant with ophthalmo-acrometic syndrome associated with bilateral anophthalmia[5].

The management of anophthalmos can be very frustrating as there is no cure for the condition. Sometimes the child may have other conditions or complications that may require monitoring. Prosthesis can be fitted to the empty sockets. Prostheses are not just for cosmetic purposes, but may also help to promote the growth of the eye socket. Conformers, which are a bit like balloons that can be expanded inside the socket, are sometimes used to further encourage the growth of the socket. In both cases, cosmesis was not a consideration. Efforts were made so that the parents will agree for prosthetic implants in both cases the but they declined. Furthermore, consanguinous marriages should be discouraged as primary anophthalmos is usually considered an autosomal recessive trait[6].

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