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CRYPTOPHTHALMOS†

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CRYPTOPHTHALMOS (hidden eye) is a very rare congenital defect originally observed by Zehender (1872) and Manz (1872). Two types of cryptophthalmos have been described, one due to complete failure in the development of the lid folds and the other to subsequent destruction and absorption of the lids. Many other congenital abnormalities have been found in association with this condition, including coloboma of the lid (Elschnig, 1914; Key, 1920; Müller, 1922; Jusefova, 1928), dermoids, cleft palate, hare-lip, facial fissure, malformed concha, and atresia of the larynx (Chiari, 1883), meningo-encephalocele (Zehender, 1872; Manz, 1872), and ventral hernia, aplasia of kidney, hoarse voice, and ear and nose defects (Jauw Soei Hah, 1950).

Dejean, Viallefont, Boudet, and Paycha (1954) reported a case with cryptophthalmos on one side and microphthalmos on the other.

The present paper presents a case of the second type of cryptophthalmos.

Case Report

A child aged 9 years, brought up as a female, was admitted to the ophthalmic section of G. M. Hospitals, Lucknow, on April 22, 1961, with bilateral cryptophthalmos. The child had been dumb and hard of hearing since birth.

Family History.—The father and mother who are consanguineous are alive and healthy. The first child, a male aged 20 years, is alive and healthy. The second, third, fourth, and fifth children, all males, died young. The sixth and seventh children, who both had right-sided cryptophthalmos with no other deformity, expired after one month. The ninth child is alive and has no congenital abnormalities.

The present case is the eighth child in the family.

There is no family history of developmental abnormality in parents.

Examination.—The eyebrows on the right side were well-developed, but on the left side there was a gap at the junction of the lateral and middle third (Fig. 1, overleaf). The lateral third was continuous with the hair of the scalp.

Both eyes were completely covered by skin, the right eyeball being bigger and more prominent. There was a small horizontal depression 5 mm. long on the lateral aspect of the skin over the right eyeball, at the upper and lower margins of which eye-lashes could be demonstrated. There was no such depression and no lashes on the left side, where all the lid structures appeared to be missing. Limited movements of the eyeballs could be seen under the skin folds. Digitally the ocular tension on the right side was high.

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Besides these local findings, a general examination of the patient revealed multiple deformities. The nose was flat and broad, with a notch at the orifice of the nostril (Fig. 2).

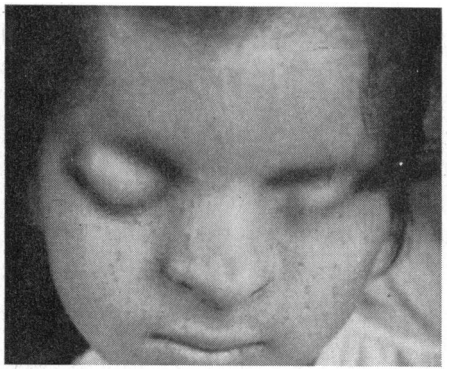

FIG. 1.—Facial appearance, showing abnormality of eyebrows and prominent right eyeball.

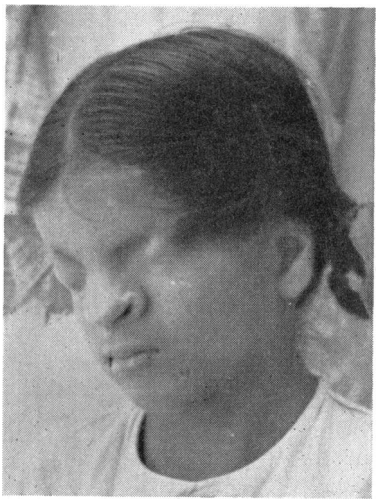

FIG. 2.—Abnormal left nostril.

There was deformity of both pinnae and atresia of the auditory canals. The tongue could not be protruded beyond the lower incisors because of a thick tongue-tie. The chest was shield-shaped, although the cardiovascular system was normal. There was an umbilical hernia. The genital organs showed gross abnormality; the labia majora were well developed, and testicular swellings could be palpated. No labia minora could be demonstrated. The clitoris looked hypertrophic, and was actually a male organ, with hypospadias on the ventral surface and a urethral opening at its root. No vaginal orifice could be seen.

The electrocardiogram and *x* rays of the orbits and skull were normal.

The Wassermann reaction and V.D.R.L. tests were negative.

Operation.—The right eye was operated on May 15, 1961, under general anaesthesia. A horizontal incision was made at the level of the depression on the lateral aspect, and the skin and subcutaneous tissue were separated. The conjunctiva was completely absent, and the subcutaneous tissue adhered to the anterior surface of sclera and cornea by thin fibrous bands. These bands were cut to expose the cornea and sclera. The cornea was ectatic and completely opaque. The right eye was enucleated and a piece of the upper lid was taken for histological study.

The left eye was similarly operated on June 6, 1961. Again the cornea was found to be opaque and adherent to the subcutaneous tissue, and the incision was sutured without further exploration.

The antero-posterior diameter of the enucleated right eyeball measured 28 mm., the horizontal 27 mm., and the vertical 26 mm. The extra-ocular muscles were not well defined, the muscle bellies being thin and inserted in an annular fashion 3 mm. behind the equator. The oblique muscles could not be differentiated. The optic nerve was thin and measured 2·5 mm. in diameter.

The saggital section of the eyeball showed that the vitreous was fluid and the inner layers intact. A small anterior chamber could be made out, but the lens, suspensory ligaments, and posterior chamber were not clearly visible.

Histological Findings.—A section through the lid showed bundles of fibro-collagenous fibres enclosing small lobules of fatty tissue and a few skeletal muscle fibres. The outer surface was lined by skin epithelium which showed well-formed sweat and sebaceous glands. There was no tarsal plate or meibomian gland (Fig. 3).

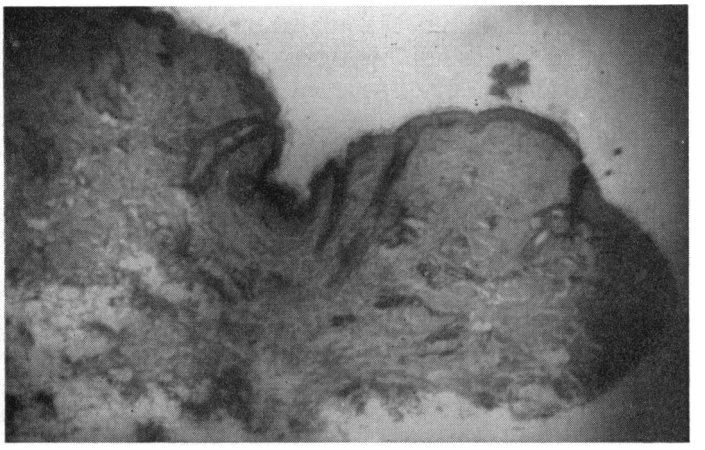

FIG. 3.—Histological section of eyelid.

A section of the eyeball showed that the cornea consisted of irregularly arranged and intertwining collagen bundles. There was no epithelial or endothelial lining and no Bowman's or Descemet's membrane was evident. The iris showed both the epithelial and pigmentary layers. The iris stroma was not well developed, and the iris was attached to the under surface of the peripheral portion of the cornea, thus reducing the size of the anterior chamber. The posterior aspect of the iris at the periphery showed irregular ciliary processes, but there was no bundle of ciliary muscles. In the region of the angle of the anterior chamber, the trabeculae and the canal of Schlemm could not be traced. The pupillary area was occupied by a small vesicle lined by a layer of columnar epithelium. The lens lamellae were not seen. The choroid, retina, and optic nerve appeared to be normally formed.

The sclera showed that the attachments of the extra-ocular muscles were slightly posterior to the equator.

Discussion

This is the first case of cryptophthalmos studied here in the past 30 years. The defect in this case was bilateral, and there were also other deformities. Undescended testes, small rudimentary penis with hypospadias, and absence of vaginal orifice led to the conclusion that the child was actually a male.

Detailed studies of the eyeball and lid revealed other abnormalities. The conjunctiva was not seen at all. The various layers of the cornea could not be demonstrated. The anterior chamber was very small. There was no trace of the trabeculae, canal of Schlemm, and ciliary muscles. The suspensory

ligaments were not developed. The lens was rudimentary and only its capsular epithelium, adherent and blocking the pupillary area, could be recognized. The extra-ocular muscles were underdeveloped and were inserted 3 mm. behind the equator. The oblique muscles could not be differentiated. The size of the eyeball was too great for a child aged 9 years and the tension was also high. This enlargement of the eyeball and raised tension (buphthalmos) were due to defective aqueous drainage.

This case belongs to the second type of cryptophthalmos as there are signs of development of the lid folds. The appearances favour an "inflammatory" aetiology, causing complete adhesion between the lid margins (ankyloblepharon) and multiple adhesions between the lids and the globe (symblepharon).

Summary

(1) A case of cryptophthalmos is reported with detailed clinical and histological studies.

(2) The patient's parents were consanguineous.

(3) The following associated general deformities were present:
- (*a*) Deformity of pinnae;
- (*b*) Atresia of auditory canal;
- (*c*) Broad flat nose;
- (*d*) Notch at orifice of left nostril;
- (*e*) Tongue-tie;
- (*f*) Umbilical hernia;
- (*g*) Gross abnormalities of the genital organs.

(4) The following associated deformities were present in the eyeball:
- (*a*) Buphthalmos;
- (*b*) Absence of trabeculae, canal of Schlemm, and ciliary muscles;
- (*c*) Rudimentary and abnormally situated lens;
- (*d*) Abnormal differentiation and insertion of extra-ocular muscles.

(5) It suggested that the term "ablepharon" should be restricted to cases of cryptophthalmos in which there is failure in the development of the lid folds. For those cases in which lid folds develop with subsequent destruction and absorption the term "total ankylosymblepharon" is proposed.

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