MICROPHAKIA*

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MICROPHAKIA or spherophakia, as a single anomaly, is very rare, but associated with ectopia lentis it is commoner (Duke-Elder, 1940). The disease results from maldevelopment of the lens and the suspensory ligaments (Mann, 1957). The ill-developed zonule is weak and does not exert its normal traction on the equator of the lens, and as a result the lens assumes a spherical rather than the normal ellipsoidal shape.

Case Report

A man aged 25 years, first seen on December 11, had experienced a sudden onset of severe pain and congestion in his right eye 5 years before. The attack had lasted for about 10 days, during which he was virtually blind. He was admitted to hospital at Srinagar, and the symptoms abated, but attacks of aching in the temporal region and pain in the eyes associated with deterioration of vision recurred frequently thereafter. With persistent attacks over a period of 12 months, the vision in the right eye was almost completely lost, and then the symptoms started in the left eye also.

Two years after the first attack he was admitted to hospital and diagnosed as a case of buphthalmos with congenital cataract in the right eye. A bluish discoloration of the sclera around the limbus was noted. Cataract extraction was done in the right eye, with iridectomy above. The family history was negative.

Examination.—Both eyeballs were prominent with marked ciliary staphyloma in the upper and lower parts of the limbus, which was unaffected in the palpebral aperture.

In the right eye the semicircular scar of the cataract operation and a wide iridectomy was seen in the upper part of the limbus. Slit-lamp examination revealed that the anterior surface of the vitreous pressed against the posterior surface of the iris. There was a scatter of fine pigment granules on the anterior surface of the vitreous in the pupillary and coloboma areas. The fundus was not clearly seen. The intra-ocular pressure was 30 mm. Hg.

In the left eye the anterior chamber was shallow—about one third of one millimetre—and the iris diaphragm was bulging into it anteriorly. The 4-mm. wide pupillary aperture was filled by the bulging anterior surface of the lens, the lower border of which was lying in front of the iris, projecting into the anterior chamber. The central area of the lens showed cataractous opacification, and biomicroscopy revealed a nuclear cataract. A crescentic area in the outer lower sector of the pupillary aperture was aphakic. The lens here appeared more spherical and smaller than normal. Through the crescentic aphakic area, the patient could count fingers at 2 metres distance. The intra-ocular pressure was 60 mm. Hg.

Treatment.—A glaucoma iridectomy was performed in the upper outer part; the equator of the lens could then be seen in the iridectomy area, and it was found that it did not

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extend to its normal distance towards the ciliary processes (Fig. 1). Only a few fibres of the suspensory ligaments were visible. The lens was found to be 5 mm. in diameter.

The upper inner edge of the lens was still behind the iris. There was a broad posterior synechia at this sector. Dusty dark-brown pigment deposits were seen on the anterior surface of the lens. The intra-ocular pressure after the iridectomy fell to 30 mm. Hg.

Progress.—After one year the patient came back with slight subjective improvement. The intra-ocular pressure was 40 mm. Hg, and was lowered to 25 mm. Hg by the use of one tablet of Diamox three times daily for 7 days. There was no complaint of pain.

The lens was extracted with a wire vectis without vitreous loss. The posterior synechia at the upper inner sector was broken without difficulty. The extracted lens had a diameter of only 5·5 mm., which confirmed the diagnosis of microphakia. Three corneal sutures were applied, and post-operative recovery was uneventful.

Result.—The anterior chamber reformed with a depth of 2·5 mm. The intra-ocular pressure was 20 mm. Hg, and the visual acuity much improved with +9 D sph.

The patient being an illiterate person, the visual acuity recorded on the dot chart was found to be 6/12 partial. Keratometry showed irregular astigmatism, the radius of curvature of cornea being 7·8 mm.; gonioscopy was not successful, however, as the contact lens did not fit over the bulging ciliary staphyloma (Fig. 2).

Comment

The cause of microphakia is supposed to be a weak zonule, so the frequency of ciliary staphyloma in this condition can be explained by the poor development of the ciliary body which gives way more easily to the increased intra-ocular pressure without leading to complete blindness.

This differs from the ciliary staphyloma seen in late stages of other types of glaucoma in adults, when it denotes complete blindness.

The cause of secondary glaucoma in these cases is the ball-valve like action of the spherical lens in the pupillary area, which causes retention of the intra-ocular fluids in the posterior segment of the eye. If this were to occur in early childhood, the eyeball would become large, because the immature scleral coat would stretch under the increased intra-ocular pressure. In such cases the condition may be wrongly diagnosed as buphthalmos.

The differential diagnosis between the two conditions is very important, because early and proper treatment gives very satisfactory results in either
case, and the necessary treatment is quite different. In buphthalmos, the cornea is big and the anterior chamber is deep. In spherophakia, the cornea is normal in size, the anterior chamber is shallow, and the posterior segment of the eyeball shows enlargement. This difference can be explained by the retention of aqueous in the posterior chamber in spherophakia, and in the anterior chamber in buphthalmos.

Summary

A case of spherophakia with secondary glaucoma and ciliary staphyloma is discussed. The case was of interest in that it simulated buphthalmos. The differential diagnosis of the two conditions is described.

REFERENCES
