Megalocornea

N.M.J.

Schweitzer

Utrecht

Author's address : Dr. N. M. J. Schweitzer, Kon. Nederl. Gasthuis voor Ooglijders, Utrecht (Holland)

A case is presented of an 8-year-old girl with megalocornea. She has a brother and sister with definite buphthalmos; they are practically blind. At the age of nine her mother was operated on for a congenital cataract. For the past three generations no parental consanguinity could be traced.

The girl had megalocornea (diameter of both corneas: 13 mm). In the angle of the anterior chamber the ciliary band was largely covered with mesodermal strands extending from the periphery of the iris towards the trabecular system, which itself was practically free of them. Schlemm's canal was not visible. In several sectors large circular vessels were seen emerging between the mesodermal strands, to disappear again in the region of the ciliary band. The chambers were not very deep. The corneas were entirely clear and Descemet's membrane was intact.

No glaucomatous symptoms were noted. The ocular tension always proved normal, as was the coefficient of the ocular outflow ($C = 0.20$; Grant's method). The visual fields were normal and the optic disc showed no evidence of cupping or pallor. There was also a normal coefficient of rigidity, 0.023 (Schiotz' 10 gram weight against applanation tonometry). The lenses were not (sub-)luxated nor the irises atrophic.

Apparently this case of megalocornea in a family with buphthalmos falls in the category of the cases presented by Kluyskens, Weekers, Malbran and others, who found megalocornea in pedigrees revealing buphthalmos. They consider megalocornea a congenital anomaly but not so far developed as to cause raised tension. To call this "incomplete glaucoma" (as Kluyskens does) would imply that at least one glaucomatous symptom is present. The term "megalocornea with angle anomaly" would separate the picture from "megalocornea vera" without pinning the patient down to the grave diagnosis of a disease, the consequence of which he has barely escaped. Whether or not the girl may show glaucomatous symptoms at any time in the future can of course not be predicted; careful observation will be necessary to recognize such evolution (vom Hofe).

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References

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Discussion

Janssens: The occurrence of megalocornea and buphthalmos has already been described in connection with gargoylism. The occurrence of buphthalmos, sometimes with cataract (in the mother of the patient), was described in connection with the syndromes of Fanconi and Lowe. In this case was there perhaps any bodily abnormality observed among the relatives?

Schweitzer: The patient and her relatives were all bodily normal and otherwise healthy.

XVaardenburg: There are two possibilities: 1. Either the other brothers and sisters have a secondary hydrophthalmos according to Peters and Rieger and this child had it in very light form; 2. Or we are dealing with primary hydrophthalmos and this girl is an abortive case, possibly brought to a halt spontaneously. Megalocornea is transmitted X-chromosomal recessively, hydrophthalmos autosomal recessively. Further, a dominant macrocornea exists.

Crone: Infantile glaucoma is ascribed to persisting embryonic tissue in the chamber angle. It is not difficult to conceive that later on spontaneous regression occurs in this tissue and that thus light cases of buphthalmos can be cured. Dr. Schweitzer's patient has no glaucoma at the moment but may certainly have had it.

Hagedoorn: I am quite willing to concur with Dr. Crone's observation. This is a difficult question and insufficiently studied. I have been struck by cases of buphthalmos that later had normal tension

although there was no trepanation filtering scar. Possibly in these cases belated development of the drainage system played a role.

Schweitzer: In my opinion
too

this case belongs in the group of primary buphthalmos and can be called an abortive form of it.