

Hypoplasia of the optic nerve

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Hypoplasia of the optic nerve is uncommon (Scheie and Adler, 1941; Somerville, 1962; Whinery and Blodi, 1963; Ewald, 1967). It is usually due to a failure of development of the ganglion cell layer of the retina and results in a small but discernible optic disc with central vessels. The disc may be cupped with a peripapillary, mottled, yellowish halo corresponding to the size of the normal disc, and the nerve fibre layer of the retina and axis cylinders of the optic nerve are absent (Scheie and Adler, 1941; Whinery and Blodi, 1963; Helveston, 1966; Ewald, 1967).

The following is a case of unilateral hypoplasia of the optic nerve in a child who was otherwise normal.

Case report

A 3½-year-old white boy had decreased vision and squint of the left eye of one year's duration. At 9 months of age the child had fallen and fractured the occipital area of the skull. There had been no neurological sequelae. At 2 years of age the mother had noticed the left eye turning in. The past history was normal except that the child had been the product of a prolonged labour and threatened abortion. The family history was negative except that the mother had epilepsy and was taking diphenylhydantoin while pregnant.

Examination

He was a well-developed well-nourished boy with a left esotropia. The visual acuity was 20/30 in the right eye, with +1.75 D cyl., axis 70°, and no light perception of light in the left eye. The corneae were both 11 mm. in diameter. The right pupil reacted well with direct light but did not react consensually when light was directed to the left eye. The left eye reacted poorly when light was directed to it but showed a brisk consensual reaction when light was directed to the right eye. The right fundus was normal with a good foveal reflex (Fig. 1). The left fundus showed a small optic nerve with a large circumpapillary crescent of granular pigmentation corresponding in size to the right disc (Fig. 2). The retinal vessels in both eyes were normal but the foveal reflex in the left eye was poor. The visual fields in the right eye were normal.

X rays of the skull and optic nerve canals were also normal, and there was no decrease in size of the left optic nerve canal. An electroencephalogram and a complete neurological examination were also normal.

Discussion

Hypoplasia of the optic nerve is rare, but when present is usually found with other abnormalities such as cyclopia, microphthalmos, anencephaly, hydrocephalus, and orbital encephalomeningocele (Ewald, 1967). Bilateral cases are usually diagnosed in infancy.

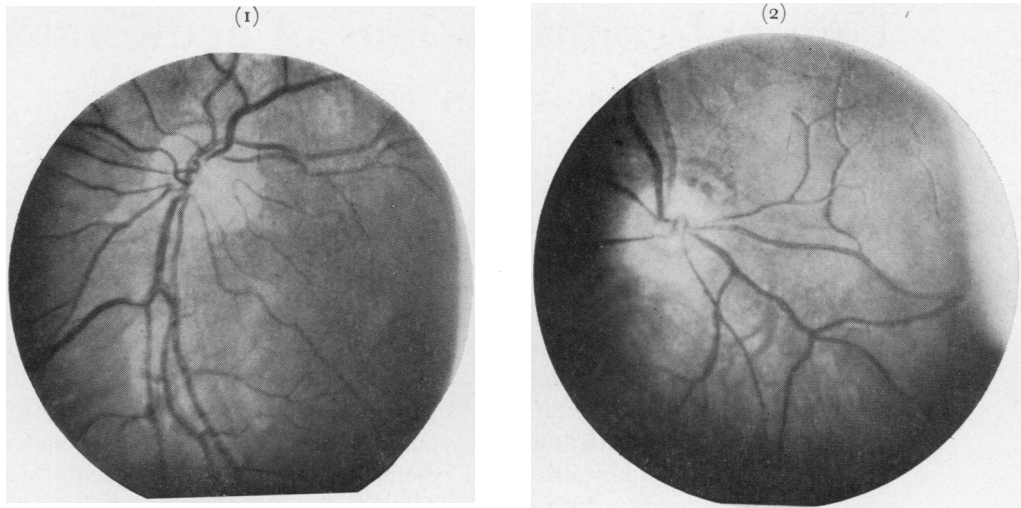


FIG. 1 *Right eye. Normal optic nerve*

FIG. 2 *Left eye. Small optic nerve with large, circumpapillary crescent of granular pigmentation corresponding to size of normal right disc*

because the blindness is obvious, but unilateral cases are usually diagnosed later in life, being regarded as cases of strabismic amblyopia.

It is usually a non-familial condition, the squint being present from infancy. The case presented above is typical of hypoplasia of the optic nerve, but neurological examination and extensive studies revealed the child to be otherwise normal. Optic atrophy of the left optic nerve is a possibility, although in such cases the disc is usually normal in size and white whereas this child's disc was extremely small, and there were no systemic or neurological signs.

Summary

A case is presented of unilateral hypoplasia of the optic nerve in a young boy who is otherwise normal.

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