CASE NOTES

CHRONIC PROGRESSIVE EXTERNAL OPHTHALMOPLÉGIA*

BY

CAHID ÖRGEN

Assistant Professor of Ophthalmology, Ankara University

CHRONIC progressive external ophthalmoplegia is a rare disease. Wilbrand and Saenger (1899) collected 32 cases from the literature, and described the condition as a distinct clinical entity, separating it from the congenital form and from the cases of external ophthalmoplegia associated with other neurological manifestations. McMullen and Hine (1921) presented three cases, but rejected one which later presented other manifestations of central nervous system involvement; according to their opinion no definite difference could be determined between the congenital and chronic types. They also annotated cases reported by Beaumont (1900; twelve cases), Ayres (1896; two cases) and Altland (1909; one case). Beaumont's cases covered four generations and were familial, never congenital, slowly progressive, never fatal. Later reports were made by Terrien (1921; one case), Calhoun (1927; annotated by Duke-Elder, 1949; number not mentioned, probably one case), Langdon and Cadwalader (1928; one case), Stone (1936; one case), Wilson (1940; one case), Fagin (1942; one case), Walsh (1947; six cases, three atypical not included), Giardini (1948; one case), Morpurgo (1948; one case), Hussain (1949; one case) and Scharf (1950; one case). Thus the number of cases to be found in the available literature was raised to 65.

Clinical Picture of the Condition

The disease presents a familial and hereditary tendency. It usually begins in childhood and slowly progressing ends in a complete external ophthalmoplegia, sparing the internal muscles. No other evidence of involvement of the central nervous system is present. The first manifestation is a slight ptosis, which usually is bilateral and shows a slow progression. In the meantime ocular movements gradually become limited, so that in 20 to 40 years external ophthalmoplegia is completely developed.

Variations in this typical clinical picture are seen in some cases. Ptosis may not begin in the second eye until long after the involvement of the first one (an interval of 25 years elapsed in the case of Fagin). Sometimes limitation of ocular movements appears first and ptosis later. Walsh observed an almost complete immobility of the eyes without ptosis in a woman whose father exhibited an almost complete external ophthalmoplegia.

When the external ophthalmoplegia is complete, the eyes assume a slightly divergent position. Some patients may complain of diplopia, but generally

* Received for publication December 22, 1952.

this symptom is missing on account of the ptosis and the slow progression. The characteristic position of the head enables the patient to see fairly well: the head is thrown back and the forehead wrinkled up.

Chronic progressive external ophthalmoplegia must be differentiated from myasthenia gravis and progressive bulbar paralysis. In myasthenia gravis the paralysis is more pronounced after exertion and disappears after a prostigmine injection. McMullen and Hine (1921), Terrien (1921), and Walsh (1947) have reported cases of chronic progressive external ophthalmoplegia in which ptosis was increased after extreme exertion; generally the influence of straining is not so marked as in myasthenia gravis, but in differential diagnosis the prostigmine test is more reliable. In chronic progressive external ophthalmoplegia, eye closure is normal, but in progressive bulbar paralysis it is difficult or even impossible because of the weakness of the orbicularis oculi.

Nuclear degeneration in the 3rd, 4th, and 6th cranial nerves was determined in cases of chronic progressive external ophthalmoplegia (Langdon and Cadwalader, 1928). As the aetiology is unknown, no aetiological treatment is possible. Crutch glasses may be used against ptosis but they are not well tolerated by many patients. Surgical therapy consists mainly of correction of the ptosis by various methods. In mild cases of lagophthalmos with normal rectus superior muscle function, the cornea does not suffer from exposure, because it turns up as the patient attempts to close the eye-lids. But in cases of chronic progressive external ophthalmoplegia, the eye-balls are fixed in the primary position of gaze and for this reason a mild lagophthalmos due to surgical treatment of ptosis is sufficient to provoke erosions and ulcerations in the cornea. Taking this peculiarity into consideration,

Walsh suggests that operation against ptosis is not indicated in these cases and reports that Spaeth advanced the same opinion. Giardini, on the other hand, corrected the ptosis of one case by performing a Blaskovics operation, but opacities of the cornea occurred later as a result of deficient closure of the eye-lids. Although this result was not satisfactory, Giardini still advises a hypocorrection of the ptosis, sufficient to permit good vision.

FIG. 1.—Before operation.

FIG. 2.—Slight divergence.

Case Report

A male patient aged 57, complained of poor vision resulting from extreme bilateral ptosis which had begun when he was 16 years old and had progressed slowly for about 10 years.

History.—Measles and varicella in childhood and malaria in youth. A first cousin, the son of his paternal uncle, presented the same drooping eye-lids. No similar ocular anomaly existed in the family.

Examination.—The physical appearance was normal. The eye-brows were constantly raised and furrows of the forehead accentuated. When speaking or fixing an object the patient tilted back his head, but yet could not see well. The width of the rima palpebrarum was 2 mm. in the right eye and 1 mm. in the left (Fig. 1). Eye closure was normal. On raising the lids with the fingers, the eyes were found to be slightly divergent (Fig. 2). Although the eyes were completely immobile, the patient was unaware of this situation until examined. When told to look to the right or left, up or down, by fixing the tip of the pen, his eyes showed an extremely slight jerky movement but no change in the direction of gaze (Figs 3, 4, 5, 6). The cornea presented an arcus senilis. Pupillary reactions to light and distance were normal. The fundi showed a tigroid appearance and the papillae a slight pallor.

FIG. 3.—Patient attempting to look to the right.

FIG. 4.—Patient attempting to look to the left.

FIG. 5.—Patient attempting to look up.

FIG. 6.—Patient attempting to look down.

Ocular Tension.—18 mm. Hg Schiötz in both eyes. Visual acuity 0/3 in both eyes and could not be corrected. The visual fields normal. Arterial blood pressure 13/8. Wassermann reaction of blood and cerebrospinal fluid negative. No pathological findings could be determined in other laboratory examinations. Nervous system normal.

Oto-rhino-laryngological examination revealed hypertrophy of the lower and middle conchae and the presence of normal reflexes at the palatum molle and posterior wall of the pharynx. Caloric stimulation of the vestibule provoked no nystagmus. Hypodermic injection of 1.5 mg. prostigmine caused no change in the ptosis.

Therapy.—As the function of the levator palpebrae superior was totally absent in our case, ptosis was hypocorrected by insertion of bands of fascia lata from lid-margin to

FIG. 7.—After operation.

frontalis muscle (Fig. 7). The patient was able to go about without difficulty after the operation. On normal closure of the eye-lids a small aperture, 1 mm. wide, was left on the right side, exposing the lower part of the cornea. The patient led a normal life for 2 months, without the use of drugs. At the end of this period a small corneal ulcer developed in the right eye, but it healed very soon when treated, and later the patient used to apply a little antiseptic ointment to the eyes every night, and this enabled him to lead a comfortable life.

REFERENCES

- ALTLAND, W. (1909). *Arch. Ophthalm., N.Y.*, **38**, 296.
 AYRES, S. C. (1896). *Amer. J. Ophthalm.*, **13**, 65.
 BEAUMONT, W. M. (1900). *Trans. ophthalm. Soc. U.K.*, **20**, 258.
 CALHOUN, F. P. (1927). *Sth. med. J., Bgham., Ala.*, **20**, 923.
 DUKE-ELDER, S. (1949). "Text-Book of Ophthalmology", vol. 4, p. 4127. Kimpton, London.
 FAGIN, I. D. (1942). *Amer. J. Ophthalm.*, **25**, 968.
 GIARDINI, A. (1948). *Riv. Oto-Neuro-Oftal.*, **23**, 181 (abstracted in *Ophthalm. Lit., Lond.*, 1948, **2**, No. 1289, p. 334).
 HUSSAIN, K. K. (1949). *Brit. med. J.*, **2**, 1392.
 LANGDON, H. and M., CADWALADER, W. B. (1928). *Trans. Amer. ophthalm. Soc.*, **26**, 247.
 MCMULLEN, W. H., and HINE, M. L. (1921). *British Journal of Ophthalmology*, **5**, 337.
 MORPURGO, F. (1949). *Atti 37 Congr. Soc. Oftal. ital.*, 1948, **10**, 397.
 SCHARF, J. (1950). *Klin. Mbl. Augenheilk.*, **116**, 654.
 STONE, T. T. (1936). *Arch. Neurol. Psychiat., Chicago*, **36**, 1380.
 TERRIEN, F. (1921). *Presse Méd.*, **29**, 937.
 WALSH, F. B. (1947). "Clinical Neuro-Ophthalmology", p. 282. Williams and Wilkins, Baltimore.
 WILBRAND, H., and SAENGER, A. (1899). "Die Neurologie des Auges", vol. 1, pp. 117–124. Bergmann, Wiesbaden.
 WILSON, S. A. KINNIE (1940). "Neurology", vol. 2, p. 1021. Arnold, London.