Clinical Cases.

DOUBLE OPHTHALMOPLÉGIA CHRONICA EXTERNA.

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The following is the clinical history of a case of chronic external ophthalmoplegia, recently observed at the Vanderbilt Clinic, the notes having been taken by Dr. Goodhart and the writer:

W. A. G., male, born in 1872, single, came to the clinic complaining of weakness in his legs and inability to use his right hand in writing. He said that after walking a short distance his legs became exceedingly fatigued. His employment as a clerk had been interfered with of late because of tremor in his right hand, so that since August last he had had to give up writing altogether. He still keeps his position, and is a weighing-clerk in a coal office.

There is nothing in the family or personal history indicative of hereditary taint or predisposition to neuroses. He has never had syphilis. He has always been temperate, even abstemious, as regards alcohol, tobacco and venery. His birth was normal, though protracted. A younger brother, the only other child in the family, is living and well. In early childhood, before the age of five years, he had diphtheria, measles and whooping-cough, from which he recovered fully. His early mental and physical development was normal. He attended school until the age of 18 years, and received a good common school education. About the age of 5 years the patient received a blow in the neighborhood of the left ear, which left a small scar. At the age of 12 years he had what seems to have been an enlarged suppurating gland below the left ear, back of the ramus of the jaw. There was a discharge for some

FIG. I.

Case of ophthalmoplegia externa at age of 11 years, showing beginning of ocular palsies.

FIG. II.

Case of ophthalmoplegia externa at present, showing marked ptosis and over-action of occipito-frontalis. Facies expressive of mental dullness.

time, and a scar remains at the site indicated. There was a discharge about the same time from the left ear. He is completely deaf in the left ear, and the drum is absent. Shortly after this a slight drooping of the left eyelid was noticed, and later of the right. Gradually, from this time on, movements of the eyes became impaired, but neither the patient nor the family can give the order of affection of the various muscles. The patient asserts, however, that he never had diplopia, and that he had never noted any particular inconvenience in the use of his eyes, save from the drooping of the lids. The patient says he was stouter in proportion as a child than at present, and that he felt much stronger generally then than now. He dates a general feeling of weakness which he has from the onset of the ptosis.

The examination reveals a slimly built, rather poorly nourished physique. He is markedly stoop-shouldered, and the head is somewhat elevated in order to assist his vision. The face is rather immobile, and the expression of countenance suggests a mild degree of mental weakness. The most striking feature, as shown in the photograph, is the marked double ptosis. The eyes would be quite closed were it not for the strong over-action of the occipito-frontalis. All of the external muscles of the eye supplied by the 3d, 4th and 6th nerves are completely paralyzed, so that the picture presented is that of ophthalmoplegia chronica externa. The irides are not involved in the paralysis, and react normally to light and accommodation. The pupils are equal. A peculiarity noticed when a strong light is thrown upon the pupils is a rapidly alternating contraction and dilatation (hippus). Dr. Carter of the Eye department reports field of vision normal, myopia, and posterior staphyloma. Vision, R 15/40—1 D 15/30. L 15/100—1 D 15/40.

With the exceptions mentioned, all of the cranial nerves are normal. Sensation of all kinds is unimpaired over the whole body. The tendon reflexes of the upper extremities

are normal, perhaps somewhat hypertypical on the right side. The patellar reflexes are hypertypical, and there is slight ankle clonus on both sides. These reflexes in the lower extremities seem to vary from time to time in degree, being more pronounced at one time than another. The abdominal, cremasteric and plantar reflexes are normal. The sphincters are normal. In the upper extremities there is no marked muscular weakness, though with the dynamometer the left hand is stronger than the right (the patient is right-handed). To simple tests the flexor muscles of the thighs appear weak. The patient has noticed quick exhaustion and weakness in his legs after short walks for a year. Although typical Romberg symptom is absent, a peculiar rapid swaying, suggestive of tremor of the trunk muscles, is observed when the patient stands with his eyes closed. The usual tests reveal considerable ataxic or intention tremor in both hands, more marked in the right. There is a similar tremor of the neck muscles exhibited upon voluntary movement of the head from side to side or when bending forward to take a drink. The speech shows a slight hesitancy, which might be regarded as an approach to syllabic utterance, but I should not call it sufficiently typical to be designated as "scanning speech." The tremor of the right hand has interfered with his penmanship only since August, 1897. He has never had any pain in his head, trunk or extremities. He has never suffered from vertigo, nausea or vomiting.

I have seen photographs of the patient at various ages. The ptosis first showed itself in mild degree at the age of 12 years. Previous to this age the appearance of the face and eyes is normal.

Remarks.—Since von Graefe, in 1856, first described the syndrome known as ophthalmoplegia externa, over 300 cases have been reported in literature, many with autopsies. The pathological process underlying the disorder is not uniform. Peripheral lesions of the oculo-motor nerves, tumors at the roots of these nerves, and growths

or softenings in the quadrigeminal region may present a similar clinical picture. Furthermore, acute or chronic nuclear lesions are frequently at the basis of an ophthalmoplegia, and to this class of cases the term *polioencephalitis superior* has been applied, because of the homology between some of the cases and cases of *poliomyelitis*. In acute lesions some *ependymitis* with punctate hemorrhages in the gray matter of the floor of the aqueduct of Sylvius has been found. The chronic form of ophthalmoplegia, to which the case just described belongs, is observed as an associated symptom in some of the chronic degenerative diseases of the nervous system, like *tabes* and *general paresis*. The chronic form is also met with in certain toxic dyscrasias (*syphilis*, *diabetes*, *diphtheria*, etc.). It is rather the rule, at least in the early stages, for the pupillary reactions to remain normal in chronic ophthalmoplegia dependent upon dyscrasias or focal lesions; whereas the internal muscles of the eye soon suffer in *tabes* and *paresis*. It is still doubtful if we have such a pathological entity as a true chronic degenerative process affecting only the nuclei of the various oculo-motor nerves, independently of any disorder elsewhere in the nervous system.

The case above reported presents certain interesting symptoms in addition to the ophthalmoplegia externa (intention tremor, increased reflexes, ankle clonus, slight hesitation in speech, and suggestive dulness of expression), which would make a multiple sclerosis more probable than any other cerebral disorder. It seems to the writer that sclerosis, with an unusual dissemination of the plaques, would explain all of the symptoms exhibited.