EXOPHTHALMOS OF CENTRAL ORIGIN *

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By exophthalmos of central origin I mean exophthalmos occurring in connexion with signs clearly pointing to an organic lesion of the central nervous system. In the short time at my disposal I can give you only a brief account of these associated symptoms and their pathological bases, and then show you the photographs of ten patients illustrating these points which I have chosen as representative, from a total of between twenty and thirty patients.

The following clinical features appear to me to be of special importance. (1) Exophthalmos may be associated: (a) with supranuclear ocular palsies, especially with paralysis of conjugate vertical movement and convergence; (b) with external ophthalmoplegia—some of these cases are etiologically obscure and exhibit bizarre defects of ocular movement; and (c) with internal ophthalmoplegia—in one case unilateral exophthalmos was associated with an Argyll Robertson pupil in the same eye; in another the reaction to light was sluggish in both eyes. (2) Exophthalmos may be unilateral or bilateral, and if bilateral it may be equal or unequal in degree on the two sides. (3) It may be associated with lid retraction, or with ptosis, either of which, again, may be unilateral or bilateral, and equal or unequal. There may be lid retraction on one side and ptosis on the other. (4) Exophthalmos may occur with or without enlargement of the thyroid. (5) It may occur with or without disturbance of sugar metabolism.

A wide range of combinations of these disturbances is encountered.

PATHOLOGY

The lesions associated with exophthalmos are pathologically varied and usually obscure; I have no post-mortem material. Encephalitis lethargica is responsible for a small number, usually with Parkinsonism. Several have high blood pressure, though in only one case is there evidence that the lesion was vascular. One patient falls into the group of internal ophthalmoplegia with absent tendon reflexes. Several can only be described as nuclear ophthalmoplegia of unknown origin. The most important single group is that in which ophthalmoplegia is associated with enlargement of the thyroid, though the relationship between the two is difficult to explain. Some of these cases have been described as myasthenia gravis associated with exophthalmic goitre. On clinical grounds it is clear that the lesion is usually situated in the mid-brain.

CASE I

The patient is a man, aged 59, who in 1927 had an illness which appeared to be encephalitis lethargica. Since then he has developed a bilateral progressive exophthalmos. He has lost conjugate ocular deviation upwards, and convergence is almost lost. There is no thyroid enlargement. His pulse rate is 72; he has lost no weight, and has an increased sugar tolerance, with a low renal threshold.

CASE II

The patient is aged 24. She began to develop Parkinsonism two and a half years ago, and now has generalized tremor and rigidity, with attacks of oculogyral spasm. She has bilateral exophthalmos, more left than right, and ptosis. There is no thyroid enlargement.

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[In another case of encephalitic Parkinsonism bilateral exophthalmos was associated with lid retraction, weakness of ocular deviation upwards, sweating, tachycardia, hyperglycaemia, and glycosuria, but the thyroid was not enlarged.]

CASE III

In 1917 the patient, a man, developed exophthalmos, tachycardia, and external ophthalmoplegia. His condition has been stationary since. He has bilateral exophthalmos and lid retraction, with partial external ophthalmoplegia. His blood pressure is 160/90; his pulse rate is 96, and he has glycosuria. His thyroid is not enlarged, and there has been no loss of weight.

CASE IV

The patient is a man aged 52, with a blood pressure of 230/120. He recently had a stroke, which left him with sensory aphasia and right homonymous hemianopia. After this he gradually developed bilateral exophthalmos with lid retraction. He has a diabetic curve in his sugar tolerance test. His thyroid is not enlarged.

CASE V

The patient is a girl, aged 23, who exhibits the syndrome of internal ophthalmoplegia, with absent tendon reflexes. In addition she has unilateral exophthalmos with lid retraction.

My last five patients all have an enlarged thyroid.

CASE VI

This patient has for six months had unilateral exophthalmos of the left eye, lid retraction, and tremor of the left hand. Her pulse rate is 108; her thyroid was not enlarged at first, but it became slightly swollen later.

CASE VII

This case, that of a girl aged 25, may be called a typical case of exophthalmic goitre, except that the patient has bilateral exophthalmos, associated with lid retraction on one side and ptosis on the other.

CASE VIII

This case, for which I am indebted to Dr. Symonds, resembles Case III in exhibiting external ophthalmoplegia with unequal bilateral exophthalmos. The ophthalmoplegia developed first, being followed by the exophthalmos, which is associated with ptosis. The thyroid is enlarged, and there is tachycardia, hyperglycaemia, and glycosuria; there has been loss of weight.

CASE IX

The patient is a woman, aged 41, who developed acutely bilateral exophthalmos, ptosis, loss of convergence, and conjugate deviation downwards, and a myotonic reaction of the pupils on accommodation. The thyroid is enlarged and nodular; there is tachycardia. The blood sugar is normal.

CASE X

A woman aged 59 began four and a half months ago to suffer from diplopia; at the same time the right eye became prominent. She feels nervous, and complains of palpitations. She has always been thin, and has lost 6 lb. in the last two years. The pupils react sluggishly to light. There is marked right exophthalmos, with bilateral lid retraction and marked weakness of upward deviation of the right eye. There is a rounded enlargement of the right lobe of the thyroid the size of a walnut; tremor of the hands, pulse rate of 100, and a blood pressure of 190/100. The blood sugar curve is normal, and the Wassermann reaction negative.

The cases I have described raise many difficult questions which I hope to discuss more fully later. All I can say now is that there seems to me to be no sharp line dividing exophthalmic goitre from certain cases of acute and chronic lesion of the nervous system, involving especially the upper part of the mid-brain.