UNUSUAL CASE OF MYASTHENIA GRAVIS*

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THE various manifestations of myasthenia gravis are numerous and the present article is intended merely to record an unusual example of the disease. The case described presented two notable features:

- (1) the length of time the disease had been present, the clinical signs being localized to the region of the eyes;
- (2) the great disability arising from the ocular abnormalities.

Case Report

A man aged 55 began to notice vertical diplopia 20 years ago. The muscle imbalance was very slight at first, and gradually became more pronounced. He was given prisms which gave temporary relief, but eventually the ocular imbalance became so great and varied in degree and type that prisms were of no assistance. Of recent years he had had great difficulty in carrying on his occupation, and driving a car had become almost impossible. He was a prominent business executive, and the eye trouble had almost forced him to the point of retiring; also, although he was not an introspective individual, the cosmetic effect of the eye lesions had caused him embarrassment at business meetings.

Examination.—In February, 1954, he was found to have ptosis of the left upper lid varying in amount from 30–80 per cent., and he said that the position of the lid varied according to the time of the day and whether he was tired or not. There was deficient elevation of both eyes, when working individually, that is with the other occluded, and when moving together. In fact, one could say there was no sursumduction at all, and the patient had to move his head to compensate for this defect. There was complete paralysis of both external rectus muscles; in the past the left eye had been more affected than the right, but now he could not move either eye outwards, and this necessitated moving his head sideways to see laterally and avoid diplopia. Diplopia both horizontal and vertical was, however, almost always present, and was so variable that accurate measurements could not be made. There was a marked esophoria and a left hyperphoria whether fixing the right or the left eye, but this did not remain constant, and tended to vary on looking upwards or downwards. The eye could not move in the direction of the paralysed muscle, so that the vertical imbalance at the extreme lateral position could not be measured. There was a refractive error, and this on correction gave normal vision. Presbyopia was normal for his age. No other abnormalities were discovered in the eyes, the pupil and ciliary functions being normal. There was no other sign of neurological disorder. Blood pressure was 150/90. The urine did not contain sugar or albumin. The Kahn test was negative. An x ray of skull showed no abnormality.

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Prostigmine.—In view of the patient's remark that the eye lesions tended to become worse towards evening and when he was tired, a provocative test with prostigmine 1·5 mg. with $\frac{1}{100}$ gr. atropine was given subcutaneously. Fifteen minutes after the injection an improvement in function of the affected muscles could be observed. After thirty minutes the patient became somewhat excited as the diplopia began to pass off, and after one hour more or less complete recovery was obtained although, naturally, ocular movements were a little sluggish. It must have been somewhat of a shock after 20 years of disability to find that his troubles had resolved so quickly. The effects of the injection lasted for some hours, and even next morning a trace of benefit remained. The injection was repeated on three consecutive evenings, the response being better each time; after the last injection the only abnormality that could be elicited was a very slight ptosis of the upper eyelid of the left eye, and the external rectus and vertical muscles appeared to have recovered their function. The patient described a feeling of stiffness in the muscles, which was only to be expected after such a long period of disuse.

Therapy.—Having made a provisional diagnosis of myasthenia gravis, regular treatment was attempted. He was put on a course of prostigmine bromide tablets (15 mg.) taking 5 a day. In one month's time he reported that he had never been so happy for years as he could now drive his car and get about normally, with no diplopia whatsoever. Examination showed perfectly straight and well-balanced eyes, with no abnormality on the cover test, 4 Worth lights, with good binocular vision. A very slight ptosis of the upper eyelid of the left eye is still discernible on close examination. Side-effects from the prostigmine therapy took the form of abdominal colic and a little diarrhoea; these were relieved by a belladonna mixture.

Follow-up.—It was not considered advisable to increase the amount of prostigmine given, and he has continued taking it for a further 4 months, with great benefit. The side-effects settled down, and he found he could do without the belladonna mixture.

It is possible that after a prolonged course of prostigmine the dosage may be reduced or even discontinued, but at the moment the patient is unwilling to do anything to prejudice the benefits obtained, and does not find it inconvenient to take prostigmine regularly.