

## BILATERAL EXTERNAL OPHTHALMOPLÉGIA.

(Ocular Myopathy.)

By P. B. B. GATENBY.

**A** MARRIED woman aged 52 complained of marked drooping of both eyelids for about ten years. For the same period she had had a squint. She thought that the condition had come on suddenly, and that in the last few years there was gradual progression of the symptoms. Otherwise she felt reasonably well, though she tended to be rather "nervous". She had never suffered from headaches nor from fits.

Her past history was uneventful except for an operation for hernia four years before. She has four children, one of whom had poliomyelitis.

On examination there was marked bilateral ptosis, with a divergent squint and almost complete lack of movements of the eyes. To compensate for the ptosis, she tended to tilt her head backwards and there was marked wrinkling of the forehead. This gave her an expression of extreme sleepiness. (See photograph.) There was also some weakness of both orbicularis oculi muscles. The pupillary reflexes were normal.

No other abnormality of the nervous system or muscles could be found. There was a mitral systolic murmur, and the blood pressure was 220/110.

X-ray investigation of the skull showed no abnormality. Chest x-ray films showed only old pleural changes and atheromatous change in the aorta. Blood Wassermann and Kahn tests negative. Urine normal.

There was no response to repeated injections of neostigmine.

*Diagnosis.* This seemed to lie between a local degenerative change in the external ocular muscles, or degeneration of the oculomotor nuclei in the brain stem. The question cannot be settled definitely without histological study of both brain and the muscles concerned, but in view of the weakness of the orbicularis oculi muscles and the normal pupils, it was considered that there was progressive dystrophy of the ocular muscles.

*Discussion.* This case is identical with the condition described and discussed by Kiloh and Nevin in 1951, and called by them ocular myopathy. They report five cases, in four of which histological proof of dystrophy of the external muscles is claimed. Also, in these cases other muscles were found to be affected clinically, such as the facial muscles, the muscles of mastication or the sternomastoids.

In an exhaustive review of about 100 papers which describe similar ophthalmoplegia, they state that there has never been any certain evidence of a lesion in the brain-stem shown histologically, and that the clinical picture has been rather that of muscular dystrophy. It is believed that the "progressive nuclear ophthalmoplegia" of the textbooks does not exist, and they are of the opinion that this ocular myopathy constitutes an important sub-group of progressive muscular dystrophy.

It gives me pleasure to thank Mr. L. E. J. Werner for examining this case and giving me help.

*Reference.*

L. G. Kiloh and S. Nevin (1951). *Brain*, 74, 2, 9.