Paralytic Exophthalmos in Chronic Progressive External Ophthalmoplegia

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Key words: chronic progressive external ophthalmoplegia, exophthalmos, mitochondrial disease

(Intern Med 51: 989, 2012)  
(DOI: 10.2169/internalmedicine.51.7119)

A 37-year-old woman was diagnosed as having chronic progressive external ophthalmoplegia (CPEO) with approximately 5 kb deletion of mitochondrial DNA fifteen years previously. Her eyeballs did not move towards any direction. She had an exophthalmos with ptosis (Picture A-C). Her eyeballs were protruded 20/17 (R/L) mm on Hertel exophthalmometer. She did not have the complication of retinitis pigmentosa. The levels of thyroid hormones and thyroid stimulating hormone were normal. Thyrotropin receptor antibody and thyroid-stimulating antibody were both negative. Computed tomography showed marked atrophy of the extraocular muscles (EOMs) (Picture D, E). In general, exophthalmos results from an increase of intraorbital volume such as in Grave’s disease. An eyeball is fixed in the orbit by the optic nerve and six EOMs. Her exophthalmos might result from the loss of supports by EOMs and this situation is not found in the early stage (1), but can occur in the advanced stage of CPEO. Although thyroid function should be examined in cases of bilateral exophthalmos, CPEO could be responsible for it if thyroid function is normal. Moreover, paralytic exophthalmos can be found in other diseases affecting EOMs such as oculopharyngeal muscular dystrophy.

The authors state that they have no Conflict of Interest (COI).

Reference