Bilateral ptosis, ataxia and areflexia—a variant of Fisher's Syndrome

Since the original description of the benign neurological syndrome of ophthamolplegia, ataxia and areflexia (SOAA) by Fisher in 1956, there has been a continuing debate as to whether the syndrome represents a variant of Guillain-Barré syndrome or a form of brainstem encephalitis. Some authors prefer the hypothesis that SOAA is a unique syndrome combining central and peripheral involvement.

A 56 year old male was admitted to University Hospital with a history of distal numbness in all four limbs, unsteadiness for four days, and severe bilateral ptosis for one day. He experienced a "flu-like" illness one week before admission. On examination, he was alert and orientated. Nasal tone in his speech was noted. Palpebral fissures were symmetrical and measured 3 mm on resting and 5 mm on maximal opening by using frontal muscles to compensate for the weakness (Fig. a, b). No limitation of extraocular movement in any direction could be detected. The pupils were 5 mm in diameter on both sides and reacted normally to light. There was no facial weakness or difficulty in swallowing. Normal convergence and Bell's phenomenon were easily demonstrated. Muscle power to all four limbs was normal. There was marked ataxia on walking and Romberg's sign was positive. Heel-to-toe test was poorly performed. Finch-to-nose and pronation-supination tests were mildly impaired. There was a generalised areflexia of the limbs. Sensory examination revealed a decrease in vibration and joint position senses in the lower limbs.

There have been a variety of clinical presentations documented which raise the possibility that the ocular problems may be supranuclear in origin, notably, a discrepancy between mild ptosis and marked external ophthalmoplegia. This patient, however, presented with isolated and symmetrical ptosis of the upper eyelid of a severe degree without limitation of extraocular movements. Eyelid ptosis is usually explained by weakness of the levator palpebrae superioris muscle due to an oculomotor nerve lesion, or by weakness of Muller's muscle secondary to involvement of sympathetic innervation, or by intrinsic disorders of the lids and their musculature. Two other possible, but less well-known causes might be "cerebral" and "midbrain" ptosis. The former may be due to failure of some control of elevation of the eyelids exerted cortically.

The latter form of ptosis may be due to the anatomical arrangement of neurons in the caudal midline of the third nerve nucleus which supply the levators of the eyelids. Clinical observation in this patient could not be explained by the involvement of infra- or supranuclear mechanisms of oculomotor nuclei. Transient, symmetrical ptosis in this patient was either due to a self-limiting and inflammatory response in the peri-aqueductal area as proposed by Meinberg or to a failure of a corticobulbar influence.

Prolongation of neck or scalp SSEP latencies in this patient with abnormal NCS were in agreement with a sensory neuropathy involving large myelinated fibres as suggested by Guiloff. It is likely that sensory polyradiculopathy plays an important role in the ataxia and areflexia of this syndrome. Based on neuro-ophthalmological observations and electrophysiological results in this patient, combined central and peripheral involvement is probable. The clinical manifestations, laboratory findings and clinical course closely resemble those reported cases of SOAA except for the ophthalmological features. Such combinations may be regarded as one of the variations of SOAA.

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Progressive supranuclear palsy as the sole manifestation of systemic Whipple's disease treated with pefloxacine

Oculomasticatory myorhythmia (OMM) with progressive supranuclear oculomotor palsy has been reported only as a cerebral complication of systemic Whipple's disease and is thus possibly a unique and pathognomonic movement disorder in this condition. Whipple's disease may be confined to the CNS so that when OMM occurs without symptoms, positive evidence for Whipple's disease may be difficult to find especially when perendoscopic and peroral jejunal biopsy specimens are normal. CSF examination and brain imaging may also be normal. In these circumstances, some authors suggest that cortical brain biopsies have a low sen-

Figure  Facial appearance of the patient at the time of admission (A and B). A) Bilateral ptosis on resting state. B) Compensatory frontalis muscles contraction on maximal opening.
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