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PTOSIS LINKED TO MYASTHENIA: CASE REPORT

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PTOSIS LINKED TO MYASTHENIA: CASE REPORT

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SUMMARY

Ptosis is the consequence of a general or an ophthalmologic pathology, whose etiological diagnosis and treatment can be complex. Eliminating life-threatening emergencies or chronic and severe diseases remain an obsession for the neurologist or the ophthalmologist. Autoimmune myasthenia gravis is a leading to evoke, especially when ptosis is fluctuating. In this disease, ptosis can be unilateral or bilateral, painless; variable during the day, aggravated by exertion, and it is frequently associated with diplopia. Ocular signs are indicative of the disease in half of the cases. We report a case of seropositive myasthenia (presence of acetylcholine receptor antibodies) in a 35 years old patient admitted for ptosis, and monitored for unexplained disorders of chewing for about a month.

INTRODUCTION

Ptosis may be linked to a general disease or an ophthalmologic pathology whose etiological diagnosis and treatment can be complex (1). Neurological ptosis is related to impairment of nerve III, either at the nucleus in the brainstem in relation with ischaemia or haemorrhage, or along its path due to compression or micro-angiopathy (1-3). Neuromuscular junction is disrupted in myasthenia gravis, which is one of the essential causes. Ptosis is characterised by its fluctuating nature and the response to specific therapy (2-5). Myopathies are responsible of bilateral and progressive ptosis. Mitochondrial myopathies, oculopharyngeal muscular and Steinert's disease are the main causes (6-10). Horner's syndrome can be responsible of a discreet ptosis (11, 12).

We report a case of ptosis associated with myasthenia gravis in a 35 years old patient, also followed for unexplained chewing disorders.

CASE REPORT

Mr. NK was a construction worker; he was admitted to the stomatology department for chewing problems for about a month. Indeed, it describes repeated and easy morsurs his tongue while eating. Sometimes with difficulty swallowing his tongue seems paralysed and unable to push the alimentaire bowl towards

the throat. This disorder of mastication was absent in the morning and very discreet afternoon. It was systematically increased late in the day when Mr NK worked. Additional tests performed (blood count, erythrocyte sedimentation rate, scanner and magnetic resonance imaging (MRI) of brain) were normal.

The appearance of a bilateral eyelid ptosis, more marked on the right, was the subject of a consultation in our department. This ptosis had the same characteristics as the trouble of chewing, that was marked in the evening after work, and was absent or discreet at rest.

Myasthenia gravis was the main suspected diagnoses; our reasoning was reinforced by the fluctuating nature of the clinical signs. For this, three clinical tests and one biological test were carried out: - *The Simpson's sign*: Day 1, we examined the patient in the morning, when the ptosis was absent. The patient was seated in front of us. We asked him to fix a target located at the top of his head. After about 30 minutes the ptosis quickly installed.

- *The test of Ice*: Day 2, we examined the patient in the evening when the ptosis was major. We applied it on the upper eyelids on both sides, just below surciliaires arcades, a piece of ice the size of a small lemon. After about 10 minutes, ptosis significantly decreased.

- *The test of Prostigmin*: Day 3; we reviewed the patient in the evening when the ptosis was major. We injected one ampoule of Prostigmin in intra-muscular. After

about 15 minutes, ptosis significantly decreased.

- *The serology* revealed the presence of anti-acetylcholine receptor antibodies.

The inadequacy of the technical platform does not allow us to search Anti-Muscle Specific antibodies Tyrosine Kinase (MuSK). The MRI of thymus was normal.

In the absence of inflammatory syndrome, the patient was put under Pyridostigmine (a derivative of the Prostigmin) without corticosteroids.

DISCUSSION

Ptosis is defined by the lowering of the upper eyelids to a defect in contraction of the levator muscle. This muscle is composed of striated and smooth muscle (Müller's muscle); innervated by sympathetic and cerebrospinal motor nerve fibers (1-4). Ptosis is caused by the alteration in any anatomical level of the specific motor pathway. The most common causes are damage of the nerve III, myasthenia gravis, various myopathies and sympathetic involvement (2-8).

Ptosis of central origin: Ischaemia leads etiologies. A bilateral ptosis can be observed in hemispheric stroke, while the alternate syndrome dominates the clinical signs in case of brainstem lesions. The painful ophthalmoplegia are particularly common in diabetic patients, the pupil is respected, and the prognosis is favorable in about three months (2, 3, 5).

Ptosis in Horton's diseases: Ptosis can be observed in this disease, regardless of blindness. It regresses within hours to weeks with corticosteroid therapy (13, 14).

Ptosis in Tolosa-Hunt's syndrome: It begins with a sudden and severe pain in the territory of nerve VI. This pain can last from several weeks to several months. It is secondarily accompanied by ptosis, ophthalmoplegia and optionally an involvement of other cranial nerves (II, VI, VII). The sedimentation speed is increased; there is lymphocytosis in the cerebrospinal fluid. MRI may show a widening of the cavernous sinus. Disorders usually regress with corticosteroids, but may recur (15, 16).

Ptosis in the ophthalmoplegic migraine: Usually the symptomatology starts in childhood by typical migraine episodes. Ptosis and ophthalmoplegia appear later. In each episode, the troubles disappear in a few days. Additional tests are normal (17-18).

Ptosis of compressive origin: Any local lesion compressing the nerve III may be responsible for ptosis, possibly painful. Imaging tests may show: tumors of various origins (meningioma, pituitary

tumors or metastasis), arterial aneurysms (posterior communicating, carotid) or sphenoid sinusitis. Lastly, nerve injuries can be directly involved (fracture, hematoma) or indirectly (temporal engagement causing ptosis, ophthalmoplegia and mydriasis) (2, 19).

Ptosis by impairment of neuromuscular junction (2-6)

1. Autoimmune myasthenia gravis

This is one of the leading diagnoses to evoke, especially when ptosis is fluctuating. Myasthenia gravis can be seen at any age, but is most common in young women. Ptosis can be unilateral or bilateral, painless, but mostly it varies outstanding day and aggravated by stress. It is frequently associated with diplopia and eye symptoms are indicative of the disease in half of the cases. There are frequently other symptoms in the cephalic territory (voice and articulation, swallowing, chewing) and often paresis of the limbs and/or axial musculature. The search of anti-acetylcholine receptor antibodies is positive in 50 to 85% of cases. In a smaller percentage of cases, MuSK test is positive. Finally, there are cases of seronegative myasthenia where neither of these antibodies are found, especially in pure eye form. The therapeutic test is often necessary for diagnosis (Edrophonium intravenous, Neostigmine subcutaneous or intramuscular, or oral Pyridostigmine). Finally, it should verify the thymus state by CT or MRI. Treatment is based on oral cholinesterase inhibitors (Pyridostigmine) usually associated with corticoids and/or immunosuppressive therapy. Infusions of intravenous immunoglobulin and plasma exchange are sometimes useful at certain stages of the disease.

2. Congenital myasthenic syndromes

These disorders are very rare, oculomotor disorders are not constant. An age of early onset and a family history should be thinking about it. Specialized teams must orient the study of different genes involved.

Botulism (20-22)

During this poisoning, ptosis is associated with ophthalmoplegia and disorders of pupillary motility. There are also problems with swallowing and phonation, motor weakness of the limbs and trunk, autonomic signs and digestive disorders.

Mitochondrial diseases (6-10)

These dysfunctions of the mitochondrial respiratory chain can affect all organs, but the damage to the ocular muscles realizes constant and progressive ophthalmoplegia. It can be isolated or associated with a host of other signs, neurological or not. Among the most frequent one can note the existence of deafness, encephalopathy, diabetes, heart disease. Disorders are permanent and slowly progressive

worsening. Heredity is mostly native, sometimes it is sporadic, and rarely inheritance is autosomal dominant or recessive. The diagnosis is supported by a hyperlactacidemia, but mainly by the histological aspects found on muscle biopsy. The mitochondrial genome studies found point mutations or large rearrangements.

Oculopharyngeal Myopathy (23, 24)

It is an autosomal dominant disorder manifesting in general after 40 years. Oculomotor involvement is dominated by ptosis, permanent and slow aggravation. The associated with swallowing disorders is constant; the proximal deficit of lower limb is inconsistent. The diagnosis is suspected on clinical presentation and family history, but confirmed by molecular genetics.

Steinert's disease (8)

This is the most common muscular dystrophies. Inheritance is autosomal dominant. Clinical signs are dominated by the distal motor deficit of four lower limb. The appearance of the face is often characteristic with a lack of facial expression, a bilateral ptosis (generally moderate) and atrophy of the temporal fossae. There are often associated signs: early cataract, cardiac disease, cognitive impairment.

Sympathetic ptosis (2, 11, 12)

Ptosis, miosis and enophthalmos are the signs of Horner's syndrome. Ptosis is linked to paralysis of smooth muscle Müller whereas skeletal muscle is still functional. It is generally unimportant. We need to find a lesion of the autonomic nervous system of the higher centers of the cervical sympathetic trunk (Pancoast Tobias' syndrome, lymphadenopathy compression lesions of the thyroid, carotid dissection) through the spinal cord (spinal cord injury, syringomyelia)

In conclusion, the ptosis is a common symptom in many diseases. Questioning and well conducted clinical examination are essential to eliminate critical emergencies and reduces the number of additional tests to a minimum.

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