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Ptosis Revealing Autoimmune Myasthenia: A Case Report and Literature Review

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ABSTRACT:
Introduction: ocular myasthenia (OM) is an autoimmune rare disease characterized by a large clinical polymorphism.
Case report: We report the case of a 32-year-old female patient, who came to our consultation with a ptosis of the left eye that appeared 6 months ago with an aggravation during the day. The ice pack test was positive. An intramuscular prostigmine test was performed and was also positive.
Discussion: Ocular disorders in myasthenia are frequent and often indicative of this autoimmune disease. The ophthalmologist is in the front line for the diagnosis of this disease, whereas the first manifestation is ocular in 90% of cases. The clinical manifestations of ocular myasthenia are characterised by a great polymorphism. Unilateral or bilateral ptosis may be associated with oculomotor disorders and may mimic some neurogenic inflammatory, tumoral or vascular pathologies that should be eliminated by further investigations. Anticholinesterase drugs are the first-line treatment for OM.
Conclusion: Auto- immune myasthenia is a life-threatening autoimmune disease. inaugural ophthalmological manifestations are very frequent. We have to evoke the diagnosis and push the investigations in front of a ptosis.

I. INTRODUCTION :

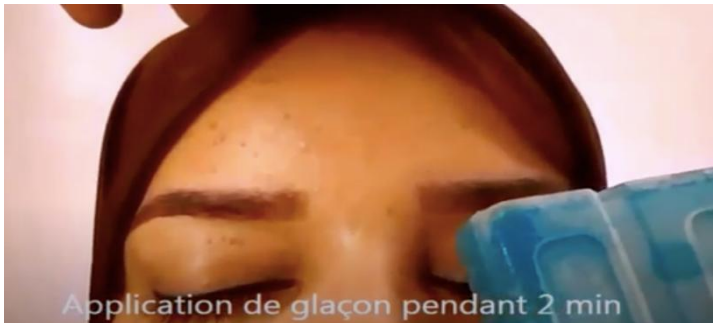
Autoimmune myasthenia is a rare disease (1), characterised by muscle weakness and abnormal fatigue on exertion. There are two main clinical forms of myasthenia: the pure ocular form resulting in ptosis, diplopia and even ophthalmoplegia, and the generalized form associating ocular signs, swallowing disorders, respiratory impairment and/or skeletal muscle deficit. We will present the observation of a 32 year old woman with ptosis revealing an autoimmune myasthenia.

II. CASE REPORT:

We present the case of a 32-year-old female patient, without any particular pathological history, who came with a slowly evolving, painless ptosis of the left eye that appeared 6 months ago. The interview revealed an aggravation during the day. Examination showed moderate unilateral ptosis of the left eye with an excursion of the levator muscle at 5 mm. The ice pack test was positive with a regression of the ptosis after 2 minutes. The visual acuity was preserved, there was no diplopia or oculomotor disorders. The Charles Bell sign was also positive. The examination of the contralateral eye was normal. The general examination did not find any general abnormalities. An intramuscular prostigmine test was performed and was positive. The anti-acetylcholine receptors antibodies test was also positive. The patient was treated by corticotherapy (0.5mg/kg/day) and azathiopirin.

Picture 1 : Moderate ptosis

Image 2 : Excursion of the levator muscle at 5 mm



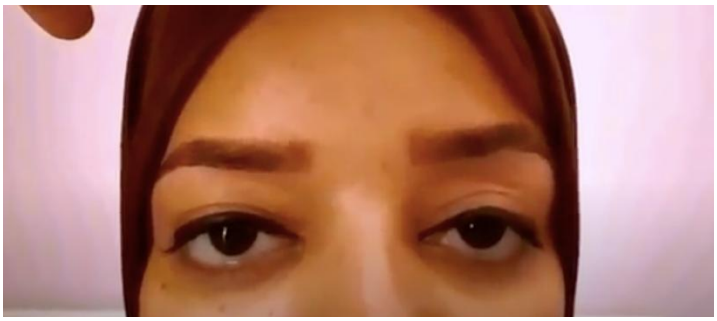


Image 3: The ice pack test shows a regression of the ptosis

III. DISCUSSION:

Auto-immune myasthenia is a life-threatening autoimmune disease due to the respiratory disorders it can cause and its possible association with a malignant thymoma. It has an annual incidence of 8 to 10 cases per million people and a prevalence of 150 to 250 cases per million. (2) The occurrence of mysthenia is influenced by gender and age: women are affected nearly three times more often than men during early adulthood (aged <40 years), whereas incidence is roughly equal during puberty and after the age of 40 years.10 After 50 years of age, incidence is higher in men. (3)

Ocular disorders in autoimmune myasthenia are frequent and often indicative of this autoimmune disease. The ophthalmologist is in the front line for the diagnosis of this disease whereas the first manifestation is ocular in 90% of cases primarily ptosis or diplopia. Two thirds of patients generalize their ocular myasthenia within two to three years of the diagnosis (4). It is important to make the diagnosis early, as some patients progress quickly to a generalized form which can be life-threatening in the short term.

The clinical manifestations of ocular myasthenia are characterised by a large polymorphism. Unilateral or bilateral ptosis may be associated with oculomotor disorders and may mimic some neurogenic inflammatory, tumoral or vascular pathologies that should be eliminated by further investigations. Ptosis is the most common clinical sign of autoimmune myasthenia. It usually has an insidious beginning, often bilateral, asymmetric or tilted, and may be isolated or associated with oculomotor disorders. In typical cases, its severity varies throughout the day and increases with tiredness. Diplopia is also a very common symptom of autoimmune myasthenia. Each oculomotor muscle may be affected individually or in association with the other muscles. The most frequently fatigued muscle in myasthenia is the medial rectus. This "simulating" aspect of ocular myasthenia justifies systematic brain and orbital imaging when it is suspected. (4) The diagnosis is clinical; the fluctuation, the worsening with fatigue and the absence of neurological systematization are characteristics of

ocular involvement. When these disorders are strictly limited to the ocular region, the diagnosis can be difficult and is made on a range of clinical and/or paraclinical grounds.

The ice pack test has a high sensitivity and specificity in the diagnosis of myasthenic ptosis. It has a higher negative predictive value and sensitivity than the prostigmine test. (5) The presence of anti-RACh antibodies is specific for autoimmune myasthenia. It is a diagnostic marker but is only found in 50% of cases of ocular myasthenia. Anti-MuSK antibodies are sometimes used to diagnose seronegative forms. The anti-striated muscle antibody test is not very sensitive but is fairly specific for myasthenia gravis. Muscle biopsy is very sensitive and specific. (6)

The frequency of thymic abnormalities in myasthenic patients requires mediastinal imaging to determine the thymic status. Thymic hyperplasia has been reported to occur in approximately 50% of myasthenic patients, while malignant thymoma is present in 10-30% of cases. (4)

Anticholinesterase drugs are the first-line treatment for OM. The response to this treatment is variable. Ptosis is often markedly improved, while diplopia is usually little changed by this medical treatment. Corticosteroid therapy is usually very effective in reducing functional symptoms in myasthenia gravis. In OM, it is proposed in case of failure of anticholinesterase drugs and when the disease is clearly invalidating. Immunosuppressive treatment, mainly with azathioprine (Imurel®) or mycophenolate mofetil (Cellept®), but also with cyclophosphamide or ciclosporin, may be proposed. Its efficacy is recognised and appears to be more or less identical to that of corticosteroid therapy. Plasma exchange or immunoglobulin infusions are not indicated in cases of OM. (6)

In addition, as anticholinesterase treatments are only partially effective on ophthalmological symptoms, almost 50% of patients will require corticosteroid therapy that is instituted gradually. Early initiation of immunomodulatory therapy may, according to some authors, reduce the risk of generalisation of the disease. (2) Even with current knowledge and available treatments, it is difficult to find the optimal treatment for each patient. Specialised diagnostic procedures and expert follow-up over time improve treatment outcomes. Close collaboration between ophthalmologists and neurologists is necessary for the ideal management of these patients.

IV. CONCLUSION:

Autoimmune myasthenia is a rare disease. The frequency of inaugural ophthalmological manifestations means that the ophthalmologist plays an important role in the early diagnosis. The diagnosis of myasthenia can be difficult because of the variability of the initial manifestations and the possible negativity of the complementary tests which does not eliminate this diagnosis.

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