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Myasthenia on a Case at Sikasso Hospital

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Abstract **Case Report**

Myasthenia gravis is aautoimmune disease which is due to a defect in the transmission of signals between the nerves and the muscles. Each year, there are 2 to 5 new cases per million people**.** We report the case of a young 35-year-old male patient seen in ophthalmology at the Sikasso hospital for ptosis and loss of visual acuity. The ptosis was improved after putting ice on the eyelids for 2 min (Ice cube test). The immunological assay for anti-MUSK (MUSK-Ab ELISA, IBL) and anti-RACH antibodies was performed. Chest CT did not detect thymus hyperplasia or thymoma, with no abnormalities of the pericardium and diaphragm. We observed an improvement in the signs after 6 months of treatment.

Keywords: Myasthenia, Ice cube test, AC anti-RACH and MUSK, Chest scanner Hôpital de Sikasso.

I. INTRODUCTION

Myasthenia gravis is aautoimmune diseasewhich is due to a defect in the transmission of signals between the nerves and the muscles. the most frequent of the neuromuscular junction but remains relatively rare [1].

Its incidence varies from 1.7 to 10.4/million/year depending on the populations studied and can reach 21/million in Barcelona.MG is a rare disease, with 8 to 10 cases per million inhabitants per year, but it exists all over the world [2].

Sheusually begins between the ages of 20 and 40and most often in men between the ages of 50 and 80, it can occur at any age [3].

This disease is clinically manifested by a fluctuating motor deficit, worsening with effort and yielding to rest. The evolution is capricious, interspersed with outbreaks that can threaten the vital prognosis by attack of the respiratory muscles [1]. When has ocular myasthenia, it is defined by an attack which is restricted to the ocular muscles beyond two years of evolution. The initial clinical manifestations involve the ocular muscles, in the form of ptosis and/or diplopia in more than 75% of cases. In half of the cases, the symptoms will not evolve and will remain limited to the eye muscles [4].

Its diagnosis remains difficult and is based on clinical criteria such as muscle fatigability, fluctuation of muscle deficit. Paraclinical examinations such as electromyography, measurement of anti-acetylcholine receptor antibodies (positive in 50% of these ocular forms of myasthenia) and the edrophonium chloride test may be lacking [4].

This during this diagnosis is most often delayed in our countries probably because of ignorance or the lack of precise diagnostic means for the differentiation of other causes (myopathies, congenital, traumatic).

Its management is multidisciplinary and calls on theanticholinesterases, oral corticosteroid therapy and/or immunosuppressive therapy.

We report a case of generalized myasthenia in a 35-year-old young subject admitted to the ophthalmology department at Sikasso hospital. With whom we have written our diagnostic and therapeutic approach.

II. OBSERVATIONS

We report the case of a young 35-year-old male patient seen in an ophthalmological consultation for progressive visual acuity loss for 3 years at the Sikasso hospital. He had no particular history or notion of trauma. On examination, visual acuity was counting the fingers at 4 meters to the right and 1/10 to the left, and 5/10 to the right and 7/10 to the left with optical correction. On physical examination, there was weight loss with generalized muscle hypotonia but especially marked at the scapular level (Fig 1), dysphonia. Ophthalmologically, bilateral asymmetric ptosis was major on the right and moderate on the left (Fig 2). The ptosis was improved after putting ice on the eyelids for 2 min (Ice cube test). The action of the levator muscle of the upper eyelid was weak at 4 mm with a limitation of movements of the eyeball. The prostigmine pharmacological test was not possible because it was not available in our structure. Examination of the anterior segment was unremarkable, ocular pressure taken in the air was respectively 14 mm Hg in the right eye and 18 mm Hg in the left eye, the fundus examined with the VOLK lens was normal in the two eyes. Faced with this symptomatology, we carried out a blood test comprising a blood count, VS, CRP, fasting blood sugar and creatinine and thyroid hormones which returned to normal. The immunological assay for anti-MUSK antibodies (MUSK-Ab ELISA,

The thoracic scanner did not detect hyperplasia of the thymus or thymoma, no anomaly of the pericardium and the diaphragm. The cranio-maxillo-facial CT scan revealed cerebral atrophy with a more marked sequelae appearance at the level of the calcarine furrows (Fig 3 & 4). The electroneuromyogram (ENMG) within the Sikasso hospital was not available. A treatment after an opinion of neurologist colleagues, an anticholinesterase namely MESTINON 60 mg due to 1 tablet 3 times a day for 2 months and prednisolone due to 1 mg per kg day for 15 days then decreasing in a decreasing way was administered. After two months of treatment and follow-up, always in consultation with the neurologists, we noticed an improvement in the symptoms. He is still under treatment and therapeutic monitoring.

Figure 1: Generalized muscular hypotonia

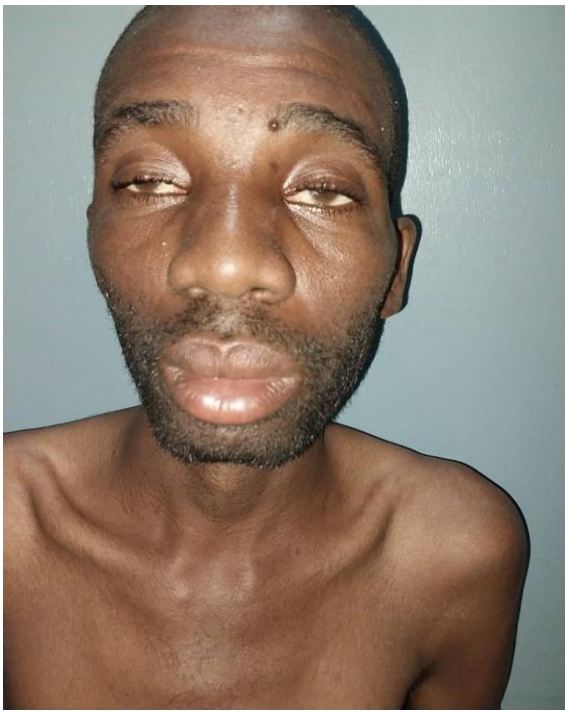

Figure 2: Increased bilateral ptosis on the right

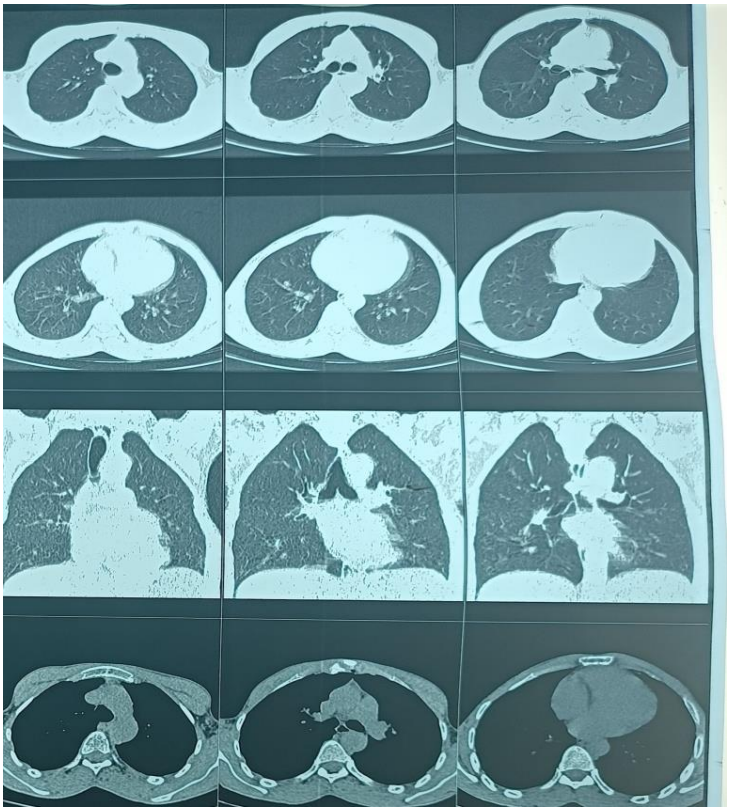

Figure 3: Thoracic CT scan with absence of thymoma

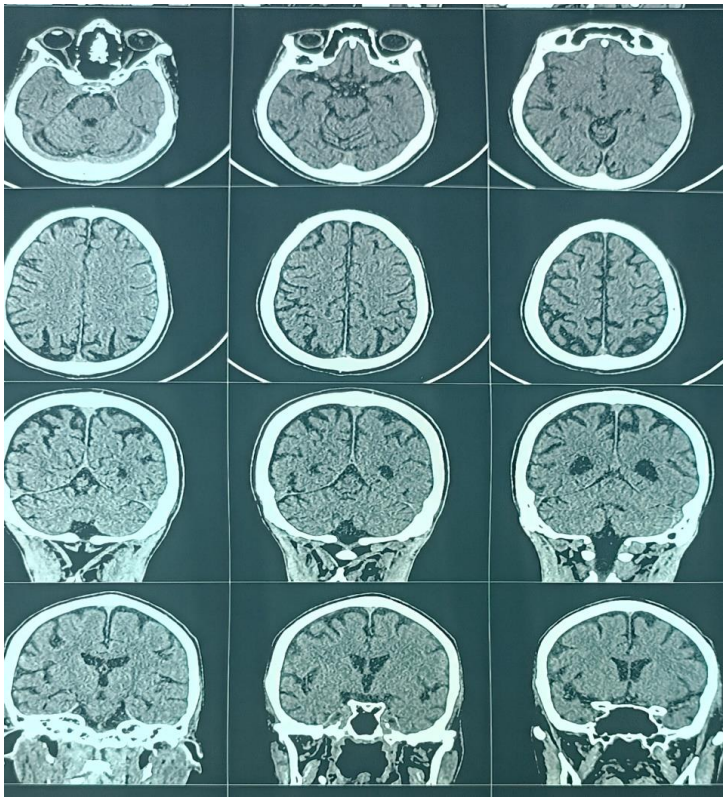

Figure 4: Craniomaxillofacial CT scan

III. DISCUSSION

Myasthenia gravis is a rare and serious autoimmune disease of the neuromuscular junction.It has a very complex clinical presentation that can simulate several pathologies, especially in its early forms [12]. She is known for the delay in her diagnosis. In our patient, the onset dates back to 3 years, he was 35 years old, which corroborates well with the study by

Djingri Labodi LOMPO which had found a more representative age group of 20 to 40 years (68%) of the cases [5].

Our age was also similar to that of Gnonlonfoun Dieudonné et al who found in a series of 6 patients with myasthenia gravis, one case in a 36-year-old man and another 33-year-old in Benin [6]. In Burkina Faso, we also found 80% of patients with myasthenia gravis with an age of onset less than or equal to 50 years [5]. However Maneh et al, in Togo, reported two cases of Juvenile Myasthenia with respectively 9 and 11 years in Togo from the same siblings with the notion of consanguineous marriage among the parents despite the rarity of family forms [7].

The sex ratio of patients is 3 women for 2 men, varies according to age group (2 women for 1 man for those under 50, 1 woman for 1 man for those over 50) [8].

Gnonlonfoun had found 4 men for 2 women in his series in Benin. On the other hand Fall M *et al*., found 4 women out of a series of 5 patients in Senegal also Djingri Labodi LOMPO *et al*., found a female predominance with 15 women (60%) and 10 men (40%) in Burkina Faso [5, 6, 9].

Santatra Ratsitohara Razafindrasata et al, in Madagascar, found a sex ratio of 0.45 in favor of women [10]. However, we cannot establish a correlation between our case, which was an isolated case, and his studies.

The reason for consultation of our patient was the decline in vision with diplopia, these signs were in 80% of cases the reasons for consultation most frequently encountered in the series of Djingri labodi in Burkina Faso [5]. The general physical examination of our patient objectified a generalized weight loss with amyotrophy of all the muscles and difficulties with physical effort (walking and lifting objects) which allowed us to classify our patient at stage III of the clinical classification of Myasthenia Gravis Foundation of America MGFA [1]. The dosage of anti-MUSK antibody (MUSK-Ab ELISA, IBL) was 0.18 IU/ml and anti-RACH was 0.2 nmol/l. These results show that it is probably a seronegative form of myasthenia gravis which represents 10 to 20% of cases of myasthenia gravis, because their absence does not allow a diagnosis of myasthenia to be excluded [12]. In Burkina Faso Djingri Labodi LOMPO found a positive AChR antibody assay in 11 out of 17 patients. In the same series of Djingri, they had found a positive result for Ab anti-MuSK in 3 patients and one patient positive for Ab anti-RACh [5].

As for the electroneuromyogram (ENMG), this was not carried out because of the absence of the device and also of the financial means achieved in the capital.

This observation of technical platform and financial means has been reported in Madagascar where socio-economic constraints and inaccessibility were the causes of the non-performance of serological examinations and ENMG [10].

The chest scanner did not find any thymoma or lung abnormality. The pericardium and diaphragm were normal in appearance.

However, Djingri labodi et al in their series of 25 patients found hyperplasia of the thymus in 12 patients, thymoma in 5 patients and a normal appearance in 8 patients [5]. Although the definitions are clear, the complementary examinations aim to confirm the diagnosis and to search for the associated pathologies (1/10 to a thymoma or an associated autoimmune disease). The dosage of these antibodies is negative in 10 to 20% of cases of myasthenia gravis. In these cases, it is all the results of the examinations (clinical and complementary) that confirm or invalidate a diagnosis of myasthenia gravis [12].

A treatment after an opinion of neurologist colleagues, an anticholinesterase namely MESTINON 60 mg due to 1 tablet 3 times a day for 2 months and prednisolone due to 1 mg per kg day for 15 days then decreasing in a decreasing way was administered. After two months of treatment and follow-up, always in consultation with the neurologists, we noticed an improvement in the symptoms.

However Dumand *et al*., reported that the effectiveness of treatment depended on certain variables, namely the earlier age of onset of myasthenia gravis, a lesser severity of symptoms at the onset, and at the maximum of the attack [11].

CONCLUSION

Myasthenia gravis remains a rare and fatal disease, especially in our countries where patients are diagnosed at advanced stages. The diagnosis of myasthenia is often clinical because biological and electrophysiological examinations are often lacking. Whatever its evolution, the treatment of myasthenia gravis is an evolutionary treatment to be reconsidered regularly.

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