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Case Report

Occurrence of Biermer's Disease After 8 Years of Follow-Up of Myasthenia Gravis: About a Clinical Case

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Abstract: The auto immune myasthenia comorbidity and Biermer disease is less documented and rarely brought in the literature. We bring back the observation of a Senegalese patient hospitalized in our department of Neurology at Fann hospital (Dakar). It concerned a patient aged 58 years followed up for auto-immune myasthenia to antibodies anti-receptors of acetylcholine Ac RACH since 8 years and having a benefit of recurrent blood transfusion with a blood group A rhesus positive. He was received on neurologic consultation for a tiredness associated to an effort dyspnea and a gastro esophageal reflux accompanied by vomiting. The interrogatory found palpitations which necessitated a hospitalization two month before. Physical examination had objective a myasthenia syndrome, an anemic syndrome on the other hand, sub icteric mucosa's were noted but no melanodermia no glossite. The rest of the physical examination was without particularity. The diagnoses of the Biermer illness was carried out in front of the anemic syndrome, the chronicity of the symptomatology without notion of fluctuation and the complementary exams having as objective a low rate of hemoglobin and a deficit in vitamin B12. The origin auto-immune of this anemia was confirmed by the immunologic test which had put in evidence a high rate of anti-bodies anti- intrinsic factor. The patient benefited from a blood transfusion then a treatment from cobalamin (for life) was also installed associated to a symptomatic management of the patient. The evolution after 6 weeks of the treatment was favorable with a complete regression of the dyspnea, vomiting and the attenuation of the effort tiredness. Myasthenia gravis and Biermer disease comorbidity has to be discussed in front of every myasthenia patient presenting clinical signs of effort dyspnea to a chronic anemia because early diagnoses of this association of Biermer illness and myasthenia gravis favors a better prognosis and not to progress to the combined degeneration of the spinal cord. The autoimmune substratum of the mechanism of this comorbidity remains to be elucidated but in all cases multidisciplinary management is necessary.

Keywords: Myasthenia, Biermer's Disease, Comorbidity, Senegal

1. Introduction

The auto immune myasthenia is a rare illness. It is associated to other auto-immune pathology in 5 to 10% of cases within this while its comorbidity with the Biermer illness is less documented and rarely brought back in the literature.

We describe here a case of a Senegalese patient hospitalized in our Neurology department of Fann Teaching Hospital of Dakar.

2. Clinical Case

It concerns a patient 58 years of age, myasthenia knows since 8 years of anti-bodies anti-receptors of acetylcholine (Ac Rach) with antecedents of recurrent blood transfusions with a blood group A rhesus positive. He was received in Neurologic consultation for a tiredness associated to an effort dyspnea and a gastro esophageal reflux accompanied with vomiting. The interrogation had revealed a notion of permanents palpitations having necessitated a hospitalization two months before his admission in our service. The physical exams had objective a myasthenia syndrome an anemic syndrome. On the other hand, sub-icteric mucosa's were being noted but no melanodermia no glossite. The rest of the physical examination was without particularity. The full blood count had put in evidence hemoglobin at 3,9g/dl, an average globular volume at 83 um^3. The rate of reticulocytes was at 30000/mm^3 (Normal value 25000 to 75000). The dosage of the vitamin B12 to 74 pmol/l (NV: 141-489). The oesogastroduodenal fibroscopy (FOGD) had showed an aspect of atrophic fundi gastric. The immunologic test, following the gastric biopsy had objectivized a rate of anti-bodies anti intrinsic factor at 14 U/ml (NV: 141-489). The myelogram showed an aspect of hypoplasia bone marrow without particular cytology abnormalities. The diagnoses of the Biermer illness was done in front of the anemic syndrome, the chronicity of the symptomatology without notion of fluctuation and the fall of hemoglobin rate and the deficit in vitamin B12. The auto immune origin of this anemia was confirmed by the immunologic test which had put in evidence an elevated rate of antibodies anti intrinsic factor. The patient benefited from a blood transfusion then a treatment from cobalamin by intravenous injection (for life). On the other hand the patient continued to follow for myasthenia an oral route treatment by pyridostigmine bromure 180 mg per day. He again benefit from omeprazole 20 mg per day. The evolution at 6 weeks of treatment was favorable with a hemoglobin at 5, 7 g/dl, a complete regression of the dyspnea, vomiting's and an attenuation of the tiredness of effort.

Auto immune comorbidity myasthenia and Biermer illnesses have to be discussed in front of every myasthenia patient presenting permanent clinical signs of effort dyspnea type associated to a chronic anemia.

3. Discussion

The pernicious anemia or Biermer disease is the most frequent manifestation of the lack in vitamin B12. This while this deficit in vitamin B12 can result from numerous causes. So the term of pernicious anemia uniquely applies to the deficit associated to a chronic atrophy gastric [1]. A lack in vitamin B12 leads to a deficit from the derived in particular from foliate, themselves coenzymes indispensable from various metabolic routes and notably from the thymidylate synthase. It's the defect to this enzyme which prevents the synthesis in DNA [2, 3], giving thus in blood cells and medullar their particular aspects [4]. The Biermer illness is part of 13 to 15% of syndrome cases multiple immune of type I and in less than 4% of auto immune syndrome cases of II and III type [5]. The Biermer illness can be associated to a certain endocrinopathy and auto-immune illness like the Hashimoto illness, the Addison illness, primary ovarian insufficiency diabetes of type I, primary hypoparathyrodis, the vitiligo and the auto immune myasthenia [6, 7].

As for the myasthenia it is a rare illness with few cases confirmed described in the African context [8], due to the effect again lake of African neurologist, to the rarity of the illness itself. Auto-immune illnesses susceptible to be associated to the myasthenia and should be systematically found are: dysthyroidy (Basedow thyroiditis) affecting 5% to 10% of patient (T4 TSH the anti-bodies of the thyroid gland), lupus, rheumatoid polyarthritis, the biermer illness and other rare illness [9]. Myasthenia is manifested at all age but two pic of incidences were described: the first about 20 years with a feminine predominance and the second after 50 years with a masculine predominance [10]. Our patient old 58 years is well in this bracket. Myasthenia at an advanced age is described in the literature [11] even above 60 years despite its reputation of being an illness of a young woman. This while the associated form begins at a young age and most often in a woman [12]. Contrary to our case of masculine sex and age close to 60 years. That could explain by late diagnostic of these systemic illnesses in current practice of our African sub-Saharan context despite the rarity of this association. In effect in a French cohort [13] of 188 patients who have attained the Biermer illness associated to other auto-immunes illnesses only 2 cases (2, 7%) were associated to an auto immune myasthenia. Whereas in large series of Biermer illness no case of myasthenia associated was rebought [7, 14], thus who testify of the rarity of this form of association of which only some cases were rebought in sub-Saharan Africa.

Nevertheless, certain large cohorts [15, 16] having brought mostly this association did not contain the immunologic test of confirmation of Biermer illness. Our patient had benefited more of a test of positive diagnoses of immunologic test of confirmation of the Biermer illness across an elevated of antibodies anti intrinsic factor at 14 U/ml (NV: inferior to 6). Even if the immunologic test is indispensable for the confirmation of diagnoses of the Biermer illness (MB), the clinical reasoning occupies an important place for the diagnostic set up of this form of association. Thus, this association has to be discussed in front of a permanent clinical decompensating in a myasthenia patient known with an

episode without any notion of symptomatology fluctuation even if we assist in an absence of dermatologic signs (melanodermy, glossite of hunter) and of the macrocytic found in a classical form of the illness. In our patient, the clinical diagnoses was mostly at the beginning guided by a permanent dyspnea associated to a tiredness always permanent despite an adequate therapeutic of her myasthenia without notion of fluctuation that which pushed us to look for an eventual association of another illness first of which the systemic illness notably the biermer illness in time with its clinical context. The lack of knowledge of such association between myasthenia auto immune and biermer illness is fatal for the patient for no benefit management is possible without a supplementation of cobalamin for life. The therapeutic of the associated myasthenia is not different from that of the isolated form [12]. Our patient benefitted from a treatment by anticholinesterase and of cobalamin (for life).

Certain author's privileged corticotherapy to that of the combination because of rechuted cases brought back [17]. The association of pathogenic mechanism of Biermer illness to myasthenia is not well elucidate but for certain author, the presence of parietal anti bodies anti cells in myasthenia patients without biermer illness suggested an auto-immune substratum common to these two affections [18].

Due to the rarity of brought back case even in great series of myasthenia patients and the state of actual knowledge on these two pathologies, a simple coincidence is not excluded in the research of the mechanism of this form of association but scientific research may be able to give an answer to this causal linkage comorbidity.

The classic evolution of Biermer's disease is combined degeneration of the spinal cord [19, 20, 21]. About our patient, the evolution at 6 weeks of treatment was favorable with a hemoglobin at 5, 7 g/dl, a complete regression of the dyspnea, vomiting's and an attenuation of the tiredness of effort.

4. Conclusion

Myasthenia gravis and Biermer disease comorbidity has to be discussed in front of every myasthenia patient presenting permanent clinical signs of effort dyspnea to a chronic anemia because early diagnoses of this association of Biermer disease and myasthenia gravis favors a better prognosis and not to progress to the combined degeneration of the spinal cord. The autoimmune substratum of the mechanism of this comorbidity remains to be elucidated but in all cases multidisciplinary management is necessary.

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