



Case Report

A Biermer's Disease Revealed by a Myogeneous Syndrome About a Rare Case in Young African Black Subject and Literature Review

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Abstract: Biermer's disease is an autoimmune disorder characterized by vitamin B12 deficiency. Neurological symptoms of B12 vitamin deficiency are polymorph. We report a case of a young black girl of 28 years old, senegalese student, consulted for walking ataxia and myalgia. The neurological examination found a myogenic syndrome of the four limbs. The electroneuromyogram showed diffuse myogenic involvement. The biological assessment found a macrocytic anemia at 112 fL with a vitamin B12 level collapsed at 74 pmol/L. The anti-parietal cell and anti-intrinsic factor were positive. Under supplementation with vitamin B12 the evolution is favorable after 8 months of treatment. Thus neurological deficiencies due to vitamin B12 deficiency are polymorphic. The dosage of vitamin B12 must be done before any neuropsychiatric symptoms that does not prove its worth.

Keywords: Biermer's Disease, Myogeneous Syndrome, Young

1. Introduction

Biermer's disease is an autoimmune disorder characterized by vitamin B12 deficiency. The causes of the deficiency are dominated by Biermer's disease and the syndrome of the vitamin B12 non-dissociation that is frequent in the elderly. Neurological symptoms of B12 vitamin deficiency are polymorph. We report the observation of young girl with a biermer's disease revealed by a myogeneous syndrome.

2. Clinical Case

Miss N. Mbaye born on 21/11/1989, student, with no specific pathological history, without a notion of consanguinity and without a notion of a particular medicinal intake, was received on 01/12/2015 for a 4 members motor deficit that had evolved since then 7 to 8 months.

The examination found:

- a) a flaccid proximal tetraparesis rated at 3/5 with myalgia
- b) a sign of the Stool and a sign of the Scarf

- c) ataxia at walking
- d) an absence of sensory disturbance

The Electroneuromyogram finds normal sensory and motor nerve conduction and at the EMG needle there were diffuse myogenic patterns without rest activities.

The muscle biopsy was not done.

In biology, there was a macrocytic anemia with a hemoglobin level of 9.7 g/dL, an average globular volume of 112 fL and a reticulocyte level of 0.7%. The level of vitamin B12 was 74 pmol/L (normal rate between 220-665 pmol/L). The metabolic biological balance was normal (Serum calcium, transaminases, creatinine, uremia, uricemia, fasting blood glucose, glycosated hemoglobin, AgHbS, anti-HCV antibody, THSUS). The infectious and inflammatory assessment (CRP, sedimentation rate, retroviral serology) was normal. The muscle enzymes: creatine phosphokinase, lactico-dehydrogenase were slightly elevated respectively to twice and one and half times normal.

The oesogastroduodenal fibroscopy showed atrophic fundial gastritis with at the histological examination:

- at the fundial level, a chronic atrophic follicular gastritis with complete intestinal metaplasia without atrophy
- at the antral level, a moderate chronic gastritis with no helicobacter pylori

The medullogram showed a rich bone marrow, marked by an important dysmyelopoiesis characterized by a maturation delay in the granular and erythroblastic lines, a presence of myeloblasts and macrocytes ((myelocytes and giant metamyelocytes) and absence of extrahematopoietic cells in favor of a vitamin deficiency (folate and / or vitamin B12).

The autoimmune balance showed negative Anti Nuclear antibodies, parietal anti-cell antibodies at 40 (normal rate less than or equal to 20) and anti-intrinsic factor antibodies at 82.56 (normal rate less than 1.2).

The diagnosis of Biermer's disease was then retained.

Under treatment with vitamin B12 injectable (1000 microgram/4 mL) due to one ampoule per day intramuscularly for one week then one ampoule per week for one month then one ampoule per month continuously, the evolution is favorable with complete motor recovery with disappearance of ataxia after 8 months of treatment.

3. Discussion

Vitamin B12 deficiency is a common condition with multiple causes, dominated by Biermer disease and vitamin B12 non-dissociation syndrome. This concept of non-dissociation of vitamin B12, described and distinguished by Carmel in 1995, gives a new face to these deficiencies especially in the elderly [1, 2]. Vitamin B12 deficiency is usually responsible for hematological abnormalities and digestive disorders. Neurological disorders are also classical [3, 4], mainly represented by combined sclerosis of the spinal cord. These neuropsychiatric manifestations have always been linked to anemia (neuroanemic syndrome), nevertheless, more and more observations of neurological disorders revealing vitamin deficiency are reported [5-6]. Megaloblastic anaemias

result from an abnormality of secondary DNA synthesis most often to a vitamin deficiency either in folic acid or in vitamin B12. In addition to haematological disorders, neurological signs are common in vitamin B12 deficiency (neurological disorders of folate deficiency are rare) and their frequency is variously appreciated. In the series of Heaton [3, 4] and those of Andr s [7], neurological disorders are reported in two patients out of three.

Biermer's disease is a rare cause of vitamin B12 deficiency. It accounts for less than 10% of the causes of vitamin B12 deficiency diagnosed in internal medicine [8]. The frequency of Biermer's disease with neuropsychiatric presentation, without anemia is variously appreciated, varying from 8.9 to 13% according to the authors [9]. It should be noted that about 1% of these patients are under the age of 50 years. In the young subject, the discovery of a Biermer disease on the occasion of isolated neuropsychiatric disorders, even MRI abnormalities is an exceptional event. The classical neurological picture of Biermer disease corresponds to that of the combined sclerosis of the spinal cord, which associates with the posterior cordal syndrome a pyramidal syndrome [3, 5]. This clinical picture, sometimes only detected by an electromyogram, accounts for 20-30% of cobalamin deficiencies and Biermer's disease with neurological presentation [3]. The other neuropsychiatric manifestations are extremely polymorphic: sensitivomotor neuropathies of more or less brutal and progressive aggravation, facial paralysis, retrobulbar optic neuritis, cerebellar syndrome, dementia... [3, 5]. There are also isolated painful forms which can be explained by an attack of the spinothalamic beam. Our observance remained unusual in that it was exceptionally described myogenic involvement in a Biermer disease. But we have asked ourselves the question of whether it is a direct complication of the disease or is it a co-morbidity. However, Regland B. et al [10] showed that the injection of vitamin B12 leads to an improvement in myalgic encephalomyelitis syndrome.

The study of the pathophysiological mechanisms at the origin of neurological and haematological disorders underlines the close link between the metabolism of cobalamin and that of folic acid. Cobalamin, as a cofactor of methionine synthetase, will transform, in the presence of folates, homocysteine into methionine [11]. The latter is transformed into S adenosylmethionine which participates in the formation of myelin basic protein [12]. At the same time, folate transformation plays an important role in the synthesis of nucleic acids of the medullary cells, oligodendrocytes, digestive cells and explains the regeneration abnormalities of these tissues during cobalamin deficiencies [4, 13]. The reasons why neurological manifestations precede haematological manifestations have so far been explained by the mechanism of folate trap [14]: the supply of folic acid would make it possible to maintain the synthesis of the nucleic acids by transformation of the folates from the dihydrofolate, to the detriment of the formation of methionine and therefore of the protein of myelin.

Methylcobalamin is required by the central nervous system

for myelin synthesis. Thus, a lack of cobalamin leads to either destruction of myelin sheaths or the incorporation of abnormal fatty acids into myelin sheaths, thereby leading to impaired neural function and/or transmissions. This may be the underlying cause of the neurological symptoms seen in vitamin B12 deficiency. However, the exact mechanism involved in myopathy due to cobalamin deficiency is not clear. In our present case, the complication of myopathy was probably related to vitamin B12 deficiency, given the absence of other causes to explain them and their cessation with vitaminotherapy.

The treatment of neurological impairment in vitamin B12 deficiency does not differ from treatment of forms without neurological impairment. Neurological recovery appears to be related to the early treatment of supplementation. The doses of vitamin B12 to be administered are not consensual. For the treatment of these neurological impairments, it is necessary to insist on the necessity of an early supplementation of vitamin B12 (complete recovery in our patient) [3, 15]. Indeed, Heaton showed that three factors were involved in the recovery: the severity of the symptoms at the time of diagnosis, the duration of the disease and the high rate of hematocrit. In his study, 57 of 121 patients with vitamin B12 deficiency with neurological signs had complete recovery within six months [5]. This was the case in our patient who completely recovered under vitaminotherapy B12.

Biermer's disease remains a rare cause of malabsorption of vitamin B12, and in even rarer cases it can be in the form of isolated neurological signs with no haematological signs that could lead to a diagnosis [16]. The neurological examination can show a complete picture of combined sclerosis of the spinal cord but often it can be limited to a central or peripheral artery but exceptionally a myogenic attack has been described. However, it is the dosage of vitamin B12 or better methylmalonic acid that will guide the diagnosis. The practice of a dosage of vitamin B12 in front of any neuropsychiatric picture seems a reasonable measure. Finally, the speed of initiation of treatment conditions the quality of neurological recovery with no sequelae in patients treated early against 50% of residual lesions in late diagnosis forms.

4. Conclusion

Neurological deficiencies due to vitamin B12 deficiency are polymorphic. They can be isolated and occur outside of any hematological context and reveal the deficiency whose etiologies are dominated by Biermer's disease and the non-dissociation syndrome of vitamin B12. The potential severity of certain neuropsychiatric complications encourages a vitamin assay in front of any neurological picture that does not demonstrate its mechanism. Early replacement therapy is the only guarantee of the prognosis of these attacks.

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