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Biermer's Disease Revealed by Normocytic Normochromic Anemia: A Case Report from the Clinical Hematology Department of Yopougon

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Abstract

Background: Biermer's disease is an autoimmune disorder, characterized by vitamin B12 malabsorption with the presence of atrophic gastropathy and various autoantibodies including antibodies to intrinsic factor. It is mainly described in Caucasian subjects in their sixties. However, it also exists in black African subjects, but occurs at a younger age. It is easily diagnosed by the association of vitamin deficiency and macrocytosis. However, various circumstances may mask the macrocytosis and thus complicate the diagnosis. The authors report the case of a patient presenting with normocytic normochromic anemia because of its rarity.

Case Report: The patient was 48 years old and was referred to a hematology consultation for investigation and etiology of a normocytic anemia of chronic evolution (Hb=8.5 g/dl, VGM=92fl, TCMH=32Pg). The diagnosis of Biermer's disease was made on the basis of the criteria of chronic anemia of insidious evolution, medullary megaloblastosis, serum vitamin B12 deficiency and the positivity of the anti-intrinsic factor antibody test.

Conclusion: This observation reminds us that a normocytic anemia does not exclude the diagnosis of vitamin B12 deficiency.

Keywords: Biermer's disease; Normocytic anemia; Vitamin B12

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Introduction

Biermer's disease is an autoimmune disease, characterized by malabsorption of vitamin B12, reversible in the presence of intrinsic factors, with the presence of autoimmune atrophic gastritis and various autoantibodies [1,2]. In Western countries, its prevalence is 9 to 17 new cases/100,000 inhabitants/year and the disease affects 0.13% to 0.20% of the population [3]. In Europe, Biermer's disease is the leading cause of megaloblastic anemia, and mainly affects Caucasian subjects, with a predominance in the elderly female subject. Indeed, 15% to 40% of patients diagnosed with Biermer's disease are over 65 years of age; 1% are under 50 years of age [2-5]. In Africa, data on the disease are scarce, but it is thought to be common in the southern part of the continent [2]: In Zimbabwe, Mukiibi reported sixteen cases in 1990; Savage in 1992, identified eighty-five cases of pernicious anemia out of 144 patients with megaloblastic anemia [6]. In West Africa, there are fewer reported cases: Akinyanju in Nigeria, reported ten, Diop in Senegal, Two cases in 1999, and Segbena in Togo, four cases in 2003 [7-9]." For all these authors, Biermer's disease is under diagnosed in black Africa because of the ancient notion of its rarity, the systematic treatment of chronic anemia without thorough investigations and the inadequacy of endoscopy technical platforms and laboratories. Its classic (historical) presentation associates neurological signs with macrocytic anemia (neuro-anemic syndrome), evolving in an insidious way, giving it the name "pernicious anemia". However, it can also occur in atypical forms, both clinically and biologically. These are forms that make the diagnosis err and ignore the disease [9-14]. In addition to the demonstration of B12 deficiency and malabsorption, the diagnosis of Biermer's disease is also based on the demonstration of autoantibodies against Intrinsic Factor (FI) which are of two types: type I antibodies, called "blockers", and type II antibodies called "precipitating" [15]. Its evolution is marked by the occurrence of complications

such as gastric polyps to be carefully monitored, more rarely one can discover gastric adenocarcinoma and finally more frequently the occurrence of multiple carcinoid tumors (4) developed at the expense of Enterochromaffin -Like Cells (ECL) [16]. Published cases are increasing and the condition is likely under diagnosed. The aim is to draw the attention of practitioners to atypical forms of Biermer's disease, the description of which is increasingly increasing in Black subjects.

Observation

The 48-year-old AK patient, a shopkeeper with an unexplored history of chronic epigastralgia, was referred to our department for exploration and etiological research of chronic normocytic normochrome anemia (Hb=8.5 g/dL, VGM=92fl, TCMH=32Pg). The anamnesis found a gradual beginning of the symptomatology that would go back about 1 year by cephalea, and dizziness she consults in a medical structure where a blood count was performed which highlighted anemia, she would have received an iron-based medication. The evolution was marked by the persistence associated with paresthesia of the extremities, skin hyperpigmentation and the persistence of anemia, which motivated a consultation in our department for better management. The clinical examination noted: a general condition preserved WHO 1, cutaneous-mucosal pallor without jaundice, a deparpillated tongue (Hunter's glossitis), epigastric sensitivity and skin hyperpigmentation (forearm, abdominal and dorsal).

The blood count noted chronic normocytic normochrome anemia (Hb=7.5 g/dL, GMV=88fl, MCDH=28Pg).

- Paraclinical examinations noted:A reticulocyte level = 25000/mm^3
- On the myelogram: Megaloblastosis with nucleocytoplasmic asynchronism
- Vitamin B12 assay 72 pmol/l
- Intrinsic factor antibodies = 52.0 u/ml
- Esogastroduodenal fibroscopy concluded an erythematous pangastric without Helicobacter pylori.
- Our patient is currently receiving specific treatment with cyanocobalamin 1,000 micrograms and proton pump inhibitor.

Discussion

Biermer's disease is rarely suspected in the face of normocytic nor-chrome anemia. This clinical case first made us look for different causes of hemolytic anemia. However, the regenerative character objectified by low reticulocytosis and the absence of splenomegaly did not plead for the classic causes of hemolytic anemia. The first cases of Biermer's disease in black subjects appeared anecdotal [17]. However, more and more cases were subsequently reported [11,18-20]. In the majority of these cases, it should be noted the insidious nature of the pathology that has been at the origin of diagnostic wanderings with fortuitous discoveries in patients admitted or initially consulting for other conditions [18,20,21]. At this level, it should be emphasized that Biermer's disease is probably under diagnosed in sub-Saharan Africa, due to a lack of technical platforms. The other reason is the lack of knowledge of certain subtleties of presentation of this condition by uninformed practitioners [20]. In the Black African subject, several striking characteristics of Biermer's disease are to be remembered, first and foremost its occurrence at a young age, compared to the age usually around 65 years in Caucasians [2,3]. Indeed, a first retrospective Senegalese work showed, between 2000 and 2007, an average age at diagnosis of Biermer's disease of 51 years in 26 patients [18]. On the other hand, a second prospective study revealed, between 2007 and 2013, an average age of 43 years in 28 cases of Biermer's disease in the same country [20]. The second characteristic is the constancy of mucocutaneous signs such as glossitis and especially palmoplantar acquired melanoderma [8,20,22]. Regarding this last cutaneous manifestation, it directs more towards adrenal insufficiency. However, it is found without further explanation in several African series and also disappears at the same time as the other signs of Biermer's disease, under well-conducted treatment [20-23]. However, melanoderma is not yet considered a pathognomonic sign of this autoimmune disease. Neurological and mucous signs (Hunter's glossitis) were present in our patient. The diagnosis of Biermer's disease was retained on the criteria of chronic anemia of insidious course, medullary megaloblastosis, serum vitamin B12 deficiency and the positivity of the search for anti-intrinsic factor antibodies whose importance in the diagnosis of pernicious anemia has been demonstrated [24]. Biologically, special presentations were noted. They consist of normocytic anemia that can be explained by the association of macrocytic and microcytic anemia by inflammation, or even microcytic related to iron deficiency or inflammatory origin [11,20,23]. Anemia, when it occurs, is consistently profound, with an average hemoglobin level of 6 g/dL [20,21]. Treatment of Biermer's disease is based on vitamin B12 administered intramuscularly as cyanocobalamin [23]. Its effectiveness is demonstrated on all clinical signs and manifestations, with however a recovery of neuropsychiatric manifestations in less than half of cases.

Conclusion

This observation reminds us that normocytic anemia does not exclude the diagnosis of vitamin B12 deficiency. The main causes of normal mean blood volume in this situation are the association of vitamin B12 deficiency with microcytic anemia (iron deficiency, hemoglobinopathy with a microcytic tendency) and the coexistence of macrocytes with fragments of erythrocytes.

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