Patient with Sickle Cell Anemia: Reflection in The Light of Medical Anthropology

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Abstract

This work aimed to approach Sickle Cell Anemia from the standpoint of medical anthropology, from the perspective of health, disease and culture, not only addressing the disease as a biological event, but also other socio-cultural aspects of the individual. This is a descriptive, theoretical-reflexive analysis-type study. Seeking to know the disease, its treatment and complications within the socio-cultural context are important to contribute to possible changes in the perspective with the individual regarding this morbidity. Having the field of public health in development, an approach beyond the technical molds for the treatment of diseases, a multidisciplinary approach is needed to meet the needs of the population, to provide patient-centered care for their social, psychological and cultural context.

Keywords: Sickle Cell Anemia; Medical Anthropology; Culture;

1. Introduction

Sickle Cell Disease has been considered a public health problem in Brazil due to its epidemiological importance in view of the high prevalence of cases [1]. The disease has its origin in Africa and was brought to the Americas as a result of the forced immigration of slaves. Due to miscegenation, this disease can be found in a large part of the Brazilian population, however, with a higher prevalence among blacks and mulattoes [2].

Defined as a set of different genotypes that are characterized by the presence of hemoglobin S (HBS), a variant alteration of normal hemoglobin (HBA), the alteration occurs by replacing the sixth amino acid of globe beta or glutamic acid by valine. Under low oxygen pressure, polymerization occurs, causing the normal shape of the RBCs to change to a scythe/middle moon shape, decreasing the life span of the RBCs [3]. This modification (HbS) causes molecules to organize into polymeric bundles when deoxygenated, giving the red blood cell an elongated and rigid form, called "scythe form" or "half-moon" [4].

There are two forms of sickle cell anemia, homozygous and symptomatic when the individual inherits the Hb S gene from his parents, he acquires the Hb SS genotype, the form of which is considered to be the most severe. In its heterozygosis and asymptomatic form, the patient presents the sickle cell trait, in which a copy of this gene is inherited from the parents, manifesting the genotype Hb AS [5]. If parents have sickle cell trait, there is a 25% probability of genetic manifestation in Hb SS and 50% in Hb AS [6].

It is important to highlight that the black population, the most affected ethnic group, is at the base of the social pyramid with the worst epidemiological, economic and educational indicators [7]. These factors contribute significantly to the poor life prognosis of people with sickle cell anemia [8]. For many years this disease was considered to be specific to black people, who came to suffer racism for the condition [9].

Although sickle cell anemia has treatment, it is a chronic disease that limits the patient to his/her daily life and makes him/her vulnerable and with biopsychosocial losses. It changes both your quality of life and that of your caregivers since it can prevent you from studying and/or working and even performing daily tasks and can generate suffering [10].

The socio-economic and educational situation, access to medical assistance and environmental conditions influence the worsening effects of the disease on the patient [11]. Researchers conducted a cohort study with patients with sickle cell anemia in the state of Minas Gerais and showed that social condition may worsen the disease and increase morbidity and mortality, which corroborates that the disease is also analyzed from political and social issues [12].

Therefore, this study aimed at reflecting on sickle cell anemia from the point of view of medical anthropology, in the perception of health, disease and culture.

2. Method

This is a descriptive, theoretical-reflexive analysis-type study. Inclusion criteria were: full articles, theses, dissertations, monographs, Course Conclusion Paper, book chapters available in the listed databases, in any language and with a five-year time frame. The exclusion criteria were: editorials, letters to the editor, abstracts, expert opinion, reviews.

Data were obtained from the Scopus, Cumulative Index to Nursing and Allied Health Literature (CINAHL), Science Direct, National Library of Medicine National Institutes of Health (PubMed) and Web Of Science databases.

The indexed descriptors were used in the *Medical Subject Headings* - MeSH: "*Anemia, Sickle Cell"; "Hemoglobin S Disease"; "Sickle Cell Anemias"; "Sickle Cell Disorders"; "Sickle Cell Diseases"; "HbS Disease"; "Anthropology, Medical"; "Medical Anthropology"*. There was a cross between them to ensure a broad search across all bases with Boolean operators OR and AND.

We found 221 articles, two of which were excluded for duplication. By applying the inclusion and exclusion criteria, 80 articles were selected. Also included in the sample by manual search, two theses and three dissertations on the subject. After reading the full text, and through eligibility, 23 primary studies were selected that addressed the topic.

3. Discussions

In order to treat sickle cell anemia under sociocultural and historical aspects, it is necessary to conceptualize

first sickle cell anemia in order to rescue its historical aspects and highlight the importance of anthropology in this area.

3.1 Sickle Cell Anemia: Evolutionary and Epidemiological Aspects

The studies of chronic diseases were approached qualitatively from the 20th century. In this century, the investigation of chronic diseases had its development based on the significance and complexity of the illness experience as a socio-cultural phenomenon [13].

Among the first studies associated to sickle cell anemia in Brazil, it was that of the physician José Martins da Cruz Jobim, who in 1835 reported in his speech about the diseases that most afflicted the lower class of the population in the city of Rio de Janeiro. Jobim reported the association between opilation and anemia, defined as intertropical hypoemia, and the perception of greater resistance to intermittent fevers by slaves coming from Africa, which was then pointed out as the protective effect of Hb S against malaria [14].

However, it was only in 1910 that the first scientific article on sickle cell disease was published. Under the authorship of doctor James Bryan Herrick, the article entitled "*Peculiar elongated and sickle-shaped red blood corpuscles in a case of severe anemia*", published in the American magazine *Archives of Internal Medicine*, demonstrated the presence of "sickle-shaped red blood cells" in a young black student from the Antilles emigrated to Chicago with severe anemia, jaundice, and intense arthralgia. Given the discovery, for years sickle cell anemia was considered a racial marker [15].

In 1922, the term sickle cell anemia was first used by Vernon Mason, and in 1927 Hahn and Gillepsie found that red blood cell sickling was dependent on low oxygen tension [16].

The 1930s was a period in which the importance of sickle cell anemia was recognized in studies in Brazilian nosology, emerging studies that addressed its clinical and pathological forms [17].

In the following decade, the goal of many researchers, such as Ernani Martins da Silva, a scientist at the Oswaldo Cruz Institute, was to correlate the frequency of sickle red cells with Brazilian racial diversity, especially to identify the degree of miscegenation [18].

Sickle cell disease was discovered, however, its inheritance was only found by the study of physician José Accioly, in 1947 in Bahia, which characterized red blood cell sickling as a consequence of an autosomal dominant inheritance [19]. But it was in 1949 that James Van Gundia Neel proved it experimentally [20].

Also in 1949, Linus Pauling and colleagues identified HbS by electrophoresis techniques, separating hemoglobins from sickle and normal erythrocytes. With this study, they pioneered the account of what they called "molecular disease" [21].

In 1954, Anthony Clifford Allison, a South African geneticist and medical scientist, did a pioneering study on genetic resistance to malaria and linked the protective effect of Hb S to disease, that is, people with sickle cell characteristics would be resistant to malaria whose agent was the highly deadly Plasmodium falciparum, which reinforced the observations already made by José Martins da Cruz Jobim in 1835 [22].

Ingran, in 1956, used the fingerprint technique (two-dimensional electrophoresis associated with chromatography) to demonstrate that HbS synthesis was caused by the replacement of glutamic acid by valine at position 6 of the β-globin chain [16].

Already in 1978, Kan and Dozy demonstrated, through molecular biology, the difference in the sequence of nitrogenous bases in the gene encoding hemoglobin beta structural chain, initiating the study of sickle cell disease through molecular biology [23].

In West, Central and East Africa there are efforts to control communicable diseases, with neglect of non-communicable diseases, hereditary blood diseases such as sickle cell disease, a consequent increase in the number of cases. Recent global estimates suggest that more than 300,000 children affected by sickle cell disease are born annually, about two-thirds of them in Africa [24]. In western Kordofan, a state of Sudan, has shown that the allelic frequency of sickle cell disease at the site is one of the highest in the world and studies on it are virtually nonexistent [25]. Even with the high mortality rate due to the disease, this condition may be a reflection of inbreeding marriages and the heterozygous advantage against malaria [26].

Data from the National Screening Program (PNTN) of the Ministry of Health, show that in Brazil are born about 3,000 children / year with sickle cell disease and 200,000 with sickle cell trait. The incidence of people with sickle cell disease is 1: 1000, and the trait in Brazil is 1:35 of live births [27].

3.2 Health, disease and culture

From the 1990s, social representations about Health and Disease have been discussed in anthropological research, aiming to address health and disease under a historical approach [28].

Culture is a set of beliefs, knowledge, behaviors, rules and values common to the social group, which provides ways of thinking and acting on what is believed [29]. The disease is subjective, whose biological process is interceded by culture, that is, the conceptions that the person has about the disease and treatment, acting and thinking come from his previous knowledge [30].

Medical anthropology is concerned with the feelings patients have about the disease, that is, based on two classic distinctions: the disease that treats the disease of the biomedical viewpoint and illness which is the experience of getting sick. These two denominations seek to address the patient's illness upon his social construction, in addition to the specific traditional and technical treatment, sustained by health professionals [31, 32].

The correlation of biology and environment as disease-causing factors is beyond the reach of culture. The modern conception of culture, accepted by most anthropologists, is more comprehensive and complex. In medical anthropology it is believed that all research, being subjective and scientific, is deepened in the culture and experience of those who interpret the results [33].

In 1944, in the field of hematology, physician Fritz Ottensooser and physician-anthropologist Ettore Biocca ensured that studies with indigenous people on blood groups for racial classification were more successful than those used by anthropology (physical anthropology) [34]. Fritz Ottensooser performed a mathematical calculation to calculate racial mixture by the frequency of blood groups, and thus, in addition to defining the concentration of indigenous black and white blood that each person retained, also indicated the degree of purity of each race [35].

On September 11, 1946, the conference “Application of the sicklemia Test to Anthropology” was held under the command of researcher Ernani Martins da Silva, which was based on his 1945 study at the Oswaldo Cruz Institute, called the “Study on sicklemia Indexes” [36]. In his research, Ernani highlighted his findings on sickle cell anemia, pointing to it as a hereditary and racial disease, especially in countries

such as Brazil, with a large contingent of black and mestizo people [37]. For him, the early detection of the disease would serve as prophylaxis of sickle cell anemia, and by the sicklemia test, it would be possible to assume whether or not the newborn would develop the disease and transmit it to his heirs.

In the 1950s and 1960s, new paths of anthropology and sociology were already being sketched out with regard to racial issues. The relationship between disease and race started to attract the attention of doctors and scholars who were thinking about Brazilian ethnic and racial formation, arousing interest in studying sickle cell anemia from an anthropological, both biological and cultural point of view [18].

Peter Fly was the anthropologist most dedicated to the study of sickle cell anemia. This researcher raised questions that sickle cell anemia could not be associated to a race, even having its origin in Africa, as it happened in the United States, where anthropologists and doctors used the disease as a racial marker, differentiating whites from blacks [9]. For this scholar, as sickle cell anemia has spread throughout the world, the disease can no longer be characterized as exceptional of the black race, but can occur in any individual.

3.3 Diagnosis and treatment: preconception and discrimination

Lopes' research identified notable discrimination against opiate-type drugs, used by patients with sickle cell anemia in a Belgian crisis, and because of this, they are now seen as chemical and opioid-dependent [38]. The dependence on this type of analgesic can happen, even though it is very well dosed and administered, however, it is also interesting the psychological follow-up for these patients. Called opio-phobia, this prejudice about the use of these drugs exists because of the lack of knowledge of health professionals in dealing with painful processes caused by the disease, and thus hinders the correct follow-up for treatment and alienates patients from the health services [39].

During the research of Ciribassi and Patil, patients reported feelings of frustration, not only because professionals did not believe in their pain, but also because they doubted the treatment with the use of opioids to cure it. Sickle cell anemia requires self-care which makes patients aware of their needs, but the doctor-patient relationship suffers stigmatization, whose professional hesitates to believe that the patient needs analgesic and, due to the hierarchy condition, the patient's request is either denied or is granted with judgments, such as the "opioid addict [40].

Thus, patients who feel illegitimate within the biomedical system refuse to proceed or seek treatment, and with the absence of the patient in the health unit, he tends to practice self-medication and self-treatment and has consequences for his health [41].

Ciribassi and Patil showed in their study that patients with sickle cell disease are often accused of using the disease itself to manipulate health professionals, with the evocative use of language and metaphor, to convince them of the pain by making it true and making it visible. However, in the face of doubt about the patients' pain, blood tests (which make the disease visible to doctors) replace the real complaint, because medicine demands a more observable and qualified reality, which makes these patients invisible when they complain and such disbelief implies in the management of care [40].

Sickle cell anemia is considered a predominant disease among blacks, who in many cases are part of the poorest groups of society, live in peripheral regions and have less access to health and education [42]. Williams and Priest have highlighted racism as a fundamental and working health cause, especially when

it comes to institutional and cultural racism, which can affect through prejudice and racial discrimination and stigma. Racism may not be the only one present in social disparities, but its presence impacts on the health of the individual and may negatively accentuate other health risk factors [43].

Another major factor to be highlighted is the significance of the disease to patients. In a survey conducted by Araújo, people with sickle cell anemia were interviewed, who pointed out morbidity as a "punishment", when they considered themselves punished due to the feeling of exclusion for being black and poor [44].

We can see that the disease reflects negatively on the patient's work activities and quality of life. However, problems such as difficulties in dealing with sickle cell anaemia, poor housing, unemployment and/or bad pay could be reduced with appropriate social and psychotherapeutic follow-up, which should be made available to these patients. In Brazil, mortality from sickle cell anemia is also estimated to be high due to the lack of implementation of public policies aimed at its approach. Patients with sickle cell disease deserve attention by the health system and through social promotion in order to reflect on improving their quality of life and life expectancy [45].

For Thornburg, Ware and each culture sees sickle cell anemia differently, and each patient and family has their individual beliefs, and so it is up to the professional team to learn to identify them. Many communities consider sickle cell anemia to be a "spell" made by witches rather than a genetic problem [46]. Others consider it a curse of some enemy, a Divine punishment, that this is the will of God or reincarnation of someone who has had the disease [47]. This way of considering the disease helps patients to think about their condition and avoid blaming themselves, in order to help them play an active role in society in coexistence with the disease [48].

Patients with sickle cell disease live with health professionals, considering that most of them have a medical follow-up, so explanations about the disease are coming from medical terms. Therefore, it is recommended that professionals, in addition to discussing diagnoses, present the disease portrayed in the social world of those who experience it. Such an approach is necessary because the patient needs both his social and cultural background and the medical environment in order to understand the disease in a clearer way and closer to his reality [49].

In view of this, it is possible to point out that issues of prejudice and stigma discriminate people and are based on stereotypes that need to be considered within the historical and social context of the person with the disease, since both consider the difficulty of society in dealing with those with differences [50].

3.4 Sickle Cell Anemia in the biocultural approach

Health and disease under biocultural analysis have biological, social, environmental, ecological and cultural characteristics, which interfere with human behavior and generate reference models [28].

The research by Lima and collaborators on quality of life evaluated 20 patients with sickle cell anemia and 40 other participants without the disease. Those with the disease perceive their quality of life as poor, with emphasis on social and physical aspects. He also pointed out that people with the disease are historically assessed as disadvantaged by the population, due to their low socioeconomic situation, which also reflects their low education, as well as the limitations that the disease itself imposes [51].

Another study by Swanson, Grosse and Kulkarni from a literature review showed that the unemployment rate among adults with sickle cell anemia is high in several countries. The same study points to an

unemployment rate ranging from 38% to 50%, and among employees, about 17% of working days suffer abstinence due to pain crises [52].

Biological vulnerability is evidenced in the clinical consequences of the disease. Intense pain, successive infections, blood transfusions, frequent hospitalizations and the need for successive treatment exemplify the materialization of these adverse conditions. There is also social vulnerability, in which the injuries caused to the physical body can reflect on the individual's mental health and, consequently, on their social relationships [53].

In a study aimed at assessing the knowledge and practices of families involved in sickle cell disease in a city in Congo, it was found that more than half of parents do not feel guilty about their child's disease. Regarding the impact on their social lives, permanent stress was the most reported condition, followed by divorce (68%). As for living with the disease, about 32% view the situation normally, 22% see it as a nightmare and 18% see morbidity as bad luck or as a will of God (28%) [54].

A study conducted in Jamaica also assessed participants' knowledge about the disease. Most said that the white blood cells attacked their red blood cells, others described that the white blood cell "sucks" the red blood cell, or that the white blood cells "eat" the red blood cells, causing an imbalance between them. In addition, much of the information is reported to have been given by health professionals, or obtained from reading materials on sickle cell anemia. In this research, as a preventive strategy to strengthen the body, participants believe that enjoying a healthy diet with iron source foods decreases the depletion of red blood cells [55].

In the same research, the patients interviewed reported that at low temperatures and prolonged periods in the water, the blood "clogs", or gets blocked causing a painful crisis. To do this, they believe that keeping the body warm makes the blood flow, and that they should also avoid rain and cold. Although medicine does not attribute sickle pain to poor circulation, doctors believe that its intensity increases on cold days and in humid conditions [55].

Dhabangi and collaborators revealed in their research in Africa that even though sickle cell anemia is well known by the population, in some communities there is no specific name for the disease, being understood in the context as loss, lack or not "having" blood. Erroneously, some patients believe sickle cell anemia was the consequence of another condition, such as poor nutrition or malaria [56].

In addition, the interviewees were asked about the clinical picture of the disease. Some reported paleness of the palmar, conjunctiva and tongue, others reported jaundice, splenomegaly, shortness of breath and poor diet, while for others the disease was caused by bad spirits [56].

A study involving 81 male and female individuals, already screened for identification of sickle cell anemia, evaluated the impact of sickle cell tracking on the population in Paris. The patients' doubts and misinformation are mainly about what is the difference between trait and disease itself, the type of hemoglobin and blood groups and lastly the Mendelian transmission. Sickle cell anemia is loaded with erroneous and stigmatizing information, as referred by a patient who believes that relating to a person, both with sickle cell trait, would not generate an individual with the disease [57].

The lack of knowledge about the disease causes embarrassment due to its appearances, such as yellowish eyes, short stature and fragile appearance. Problems arising from difficulties related to the provision of financial support and sex life, cause depression in these patients [53].

Therefore, Canesqui's considerations can be reiterated "[...] to approach the socio-cultural dimension of long-term illnesses means looking at the subject living with a condition that accompanies him everywhere [...]"[31].

4. Conclusion

People with sickle cell anemia not only face a biological condition, but the disease is also part of their life, promoting various changes that impact their emotional and social state. They impact on the life of the sick person and the family, and are also a challenge for public programs and policies.

The suffering of patients with the disease is experienced as a result of situations such as late diagnosis, misinformation about the disease, dependence and difficulty in accessing health services, prejudice due to the use of medication, body changes, lack of employment, loss of freedom, depression, weakening of family and social bonds.

Living with sickle cell disease requires the ability to meet the challenges that the disease itself imposes. Seeking to know the disease, its treatment and complications within the socio-cultural context are important to help change the individual's perspectives on this morbidity. Understanding the disease and its changes is of great importance for the individual to understand his life history and obtain biological control of his body in the face of this chronic disease.

Health professionals should be prepared from initial training to deal with situations involving socio-cultural aspects and issues of vulnerability, since it is not only the biomedical and technical part that contribute to the improvement of the patient, but also consider the socioeconomic and biocultural context that this patient experiences. For often what makes treatment difficult is the stigmata he suffers, and the vulnerable conditions in which he finds himself.

Finally, medical anthropology, as a reflexive and critical area, should be a compulsory subject in undergraduate health courses, since many diseases are considered public health problems. Having the field of public health in development, an approach beyond the technical molds for the treatment of diseases, a multidisciplinary approach is needed to meet the needs of the population, to provide patient-centered care for their social, psychological and cultural context.

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