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Dental Manifestations of SSFA2 Homozygous Sickle-Cell Anaemia: Clinical Data From A Cohort of Sixty Patients Monitored in A Referral Centre in Abidjan (Côte D'Ivoire)

Gnagne-Koffi Y[1], Djolé SX[1] and Aidara AW[2*]

[1]Department UFR of Odonto-Stomatology, Félix Houphouët Boigny University, Cocody (Abidjan, Ivory Coast)
[2]Faculty of Medicine Pharmacy and Odontology, Department of Odontology, Conservative Dentistry Division Cheikh Anta Diop University, BP 5005 Dakar, Senegal

[*]**Corresponding Author:** Aidara AW, Faculty of Medicine Pharmacy and Odontology, Department of Odontology, Conservative Dentistry Division Cheikh Anta Diop University, BP 5005 Dakar, Senegal, Ph: + 221 77 454 24 55, E-mail: wakha.aidara@ucad.edu.sn

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Abstract

Introduction: Sickle-cell anaemia is a real public health problem in Côte d' Ivoire. The objective of this study is to identify the dental manifestations of patients suffering from homozygous sickle-cell anaemia (SSFA2) in order to propose multidisciplinary management of this condition.

Patients and Methods: The study took place in the Clinical Hematology Department of West Africa located within the Yopougon Hospital and University Center (Abidjan - Côte d'Ivoire). A retrospective analysis of the records of homozygous sickle cell disease patients with SSFA2 followed by oral maintenance and examination were performed.

Results: A total of 60 patients made up our sample. The first dental manifestations in homozygous sickle-cells are represented by pain (75%) during sickle-cell attacks. There is localized pain on apparently healthy teeth. Dental pathologies are predominantly irreversible pulpopathies (43.3%) and the red blood cell deficiency appears to increase susceptibility to tooth decay.

Discussion: Dental caries and ischemia caused by the vaso-occlusive crisis represent the etiologies of dental pain in homozygous sickle cell disease (SSFA2). This painful symptomatology tends to disappear quickly with the recurrence of hemolytic attacks leading to pulp necrosis.

Conclusion: Comprehensive multidisciplinary management involving systematic odontological consultation, such as ophthalmologic consultations, is essential.

Keywords: *Homozygous Sickle-Cell Anaemia (Ssfa2); Sickle-Cell Attacks; Dental Manifestations*

Introduction

Sickle-cell anaemia or sickle cell anaemia is due to a genetic mutation that results in the substitution of the 6th Aminated Acid in the? chain of globine (glutamine) with valine [1]. It is the most common hemoglobinopathy, and epidemiological data reveal a higher prevalence (above 10%) between the 15th parallel of north latitude and the 20th parallel of south latitude called "Lehmann's Sicklemic Belt" [2]. SSFA2 homozygous sickle-cell anaemia, a major form of sickle-cell disease, is a chronic hemolytic anaemia interspersed with hemolytic attacks and often complicated vaso-occlusive attacks of severe bacterial infections [3,4]. Pioneering work has shown the impact of the disease on skeletal and mandibular bone alteration, and reported cases have shown that the oral sphere may be the site of various complications [5,6]. The dental organ is made up of a specialized connective tissue called dental pulp and three mineralized hard tissues, enamel (at crown level) and cement (at root level), which cover the dentine, the main mass of the tooth. Dental pulp is made up of a nervous, vascular and lymphatic network, which ensures sensitivity, the supply of nutrients and immune defence [7]. This network allows communication with surrounding support structures (the periodontium) such as the alveolo-dental ligament and alveolar bone [8]. The first studies on the manifestations of sickle-cell anaemia at the dental level showed the

presence of sickle-cell anaemia in pulp vessels 24 to 96 hours after the sickle-cell attack and several investigations have shown the presence of necrotic pulp teeth in sickle-cell patients [9,10]. In Côte d' Ivoire, the prevalence of haemoglobin S is high, as in sub-Saharan countries [3] and makes the disease a real public health problem. To this end, there is one of the largest regional reference centres for the diagnosis, treatment and follow-up of patients. To this end, there is one of the largest regional reference centres for the diagnosis, treatment and follow-up of patients. The objective of this study is to identify the dental manifestations of patients suffering from SSFA2 homozygous sickle-cell anaemia in order to propose multidisciplinary management of this condition.

Patients and Methodology

The study took place in the Clinical Hematology Department of West Africa located within the Yopougon Hospital and University Center (Abidjan - Côte d' Ivoire). It was carried out over two months in two main phases, including a retrospective analysis of patient files, which consisted of a review of patient files followed by a prospective investigation combining interviews and oral examinations.

Prospective survey

The medical records of sickle cell patients with hemoglobin phenotype have been consulted. Information relating to the discovery of the pathology, hematological analyses, periodicity and type of seizure (hemolytic or vaso-occlusive) was recorded.

Oral Interviews and Examinations

Each patient was asked about the annual frequency of onset of sickle-cell attacks and the association of dental pain with the occurrence of these attacks. The intensity of the pain was assessed according to the Analog Visual Scale (AVS) with "0" for no pain,[1-4] for mild pain,[5-7] for moderate pain,[8-9] for severe pain and "10" for very severe and insomniac pain. Subsequently, the oral examination was carried out in search of decayed teeth in order to make the associated diagnoses.

Statistical analysis

The data collected was processed by the EPI Info version 6.01 software. Statistical analyses were performed using the Chi2 test and the Kruskall-Wallis non-parametric test was used to compare the means between qualitative and quantitative variables. The significance threshold is set at 5% for observed differences.

Results

Sixty patients aged 12 to 40 years old, 56.7% of whom were women and 43.3% of whom were male, were selected for complete records and consent to participate in the study. A large majority of patients (85%) had been diagnosed with SSFA2 homozygous sickle-cell anaemia before the age of 12 years (Table I). Blood counts of patients are characterized mainly (85%) by erythrocyte deficiency with white blood cell (65%) and platelet (62%) levels higher than normal (Table II). With respect to the frequency of seizures, records data were superimposed on patient claims. During these attacks, 75% of the latter confirmed that they had experienced dental pain and at the request of the localization, 26.7% of them indicated apparently healthy teeth, free of carious lesions, traumas, periodontal injuries and previous dental care (Table III). The majority of patients (55.6%) had fewer than three seizures per year, and 44.4% of patients had pain on decayed teeth and 11.2% had pain on apparently healthy teeth (Table IV). Vaso-occlusive seizures are the largest in the sample (46.7%) and dental pain during these seizures involve 37.8% of decayed teeth and 8.9% of apparently healthy teeth (Table V). Oral examination revealed that 65% of the patients in the sample had carious lesions, but this proportion was mainly due to patients with erythrocyte deficiency, i. e. 60% (Table VI). The majority of dental manifestations are characterized by carious lesions of dentin (18.3%), pulsed inflammations with painful symptomatology of decay-free teeth (20%) and peripheral lesions at the top of the dental roots (periapical lesions) of decay-free teeth (15%) (Table VII).

Sample characteristics		n (%)	N (%)
Gender	Female	34 (56.7)	60 (100%)
	Male	26 (43.3)	
Age bracket	[10-20] years	34 (57)	60 (100%)
	[21-30] years	21 (35)	
	[31-40] years	5 (8)	
Diagnostic period for sickle cell disease	Before 12 years	51 (85)	60 (100%)
	After 12 years	9 (15)	

Table I: Demographic characteristics of the sample

Hematogram		n (%)	N (%)
Erythrocytes	Normal	9 (15)	60 (100%)
	Abnormal (lower)	51 (85)	
Leukocytes	Normal	21 (35)	60 (100%)
	Abnormal (upper)	39 (65)	
Platelets	Normal	23 (38)	60 (100%)
	Abnormal ((upper)	37 (62)	

Normal values: Erythrocytes [Female 4-4.5.106/mm^3; Male 4.5-6.10 6/mm^3] Leukocytes[4-10.10^3 / mm^3] Platelets[150000- 400000 / mm^3]
Table II: Distribution of Patient Hematologic Parameters

Painful manifestations		n (%)	N (%)
Dental pain during seizures	yes	45 (75)	60 (100)
	No	15 (25)	
Status of tooth with painful symptomatology	Decayed tooth	34 (75.4)	45 (100)
	Apparently healthy tooth	11 (24.6)	

Table III: Patient distribution by occurrence of dental pain during sickle-cell attacks and their location

Frequency of seizures	Pain localization		Total N (%)
	Decayed tooth N (%)	Apparently sound tooth N (%)	
< 3	20 (44.4)	5 (11.2)	25 (55.6)
[3 – 5]	6 (13.3)	5 (11.1)	11 (24.4)
> 5	8 (17.7)	1 (2.3)	9 (20)
Total	34 (0)	11 (0)	45 (100)

Chi-square = 3.76 ddl = 1 p = 0.001 (significant test)
Table IV: Patient distribution between frequency of seizures and teeth with painful symptoms

Type of seizure	Pain localization		Total N (%)
	Decayed tooth N (%)	Apparently sound tooth N (%)	
Hemolytic	6 (13.4)	4 (8.8)	10 (22.2)
Vaso-occlusive	17 (37.8)	4 (8.9)	21 (46.7)
Mixed	11 (24.4)	3 (6.7)	14 (31.1)
Total	34 (75.6)	11 (24.4)	45 (100)

Chi-square = 1.71 ddl = 2 p = 0.4252 (not meaningful test)
Table V: Patient distribution between type of seizure and teeth with painful symptomatology

Number of red blood cells /mm3	Patients		Total N (%)
	Without cavity N (%)	With cavity N (%)	
[0-3.10^6]	15 (25)	36 (60)	51 (85)
[4 -6.10^6] VN*	6 (10)	3 (5)	9 (15)
Total	21 (35)	39 (65)	60 (100)

*Normal value Khi 2 = 3.17 ddl = 1 p = 0.0074 (significant test)
Table VI: Distribution of patients between erythrocyte counts and the presence of decayed teeth

Dental manifestations	Associated diagnoses	n (%)	N (%)
Dentinal Manifestations	Dentine carious lesion	11 (18.3)	14 (23.3)
	Hypomineralizations	03 (5)	
Pulp pulses	Irreversible pulpopathy with pain without carious lesion	12 (20)	26 (43.3)
	Irreversible pulpopathy with pain with carious lesion	03 (5)	
	Chronic irreversible pulpopathy with carious lesion	05 (8.3)	
	Irreversible pulpopathy with pulp necrosis and carious lesion	06 (10)	
Periapical Manifestations	Acute acute carious apical periodontitis	01 (1.7)	10 (16.7)
	Chronic apical periodontitis of carious origin	09 (15)	
Patients free of dental pathology (carie, trauma, periodontal disease)		10 (16.7)	10 (16.7)
Total		60 (100)	60 (100)

Acute apical periodontitis: ligament and bone damage confined to the tip of the root with pain; Chronic apical periodontitis: ligament and bone damage confined to the tip of the root without pain
Table VII: Types of manifestation by dental tissue

Overall, 16.7% of patients are free from all dental manifestations, with the latter represented 43.3% of the patients being free from all dental manifestations.

Discussion

Systemic and loco-regional manifestations of sickle-cell anaemia

Sickle cell disease is described primarily in black subjects [3]. Up to the 5th - 6th month postpartum, the pathology remains asymptomatic as red blood cells contain high levels of fetal haemoglobin, blocking the falcification process. It is then very severe, marked by a critical phase called the vaso-occlusive crisis. It is the most common syndrome and is essentially represented by pain. This pain sits on the hands and feet "hand foot syndrome" and is accompanied by fever in infants. In children and adolescents, the pain is abdominal, caused by visceral infarction (mesenteric, spleen, liver) and is accompanied by fever [4,11]. These symptoms are at the origin of the consultations, resulting in a higher proportion of patients identified as suffering from the pathology as early as childhood in this study.

Etiopathogenic mechanisms of dental pain during sickle cell attacks

The etiology of dental pain in sickle cell disease is explained by ischemia caused by the vaso-occlusive crisis. In fact, it is due to the loss of elasticity of deformed red blood cells, which results in obstruction of vascular light by sickle cell anaemia creating ischemia [1]. This explains the low or moderate pain, contrary to the intense pain encountered during pulp aggression. Indeed, in this type of process, it is the synergistic intervention of inflammation mediators that will cause vasodilation, and the increase in pulp pressure associated with the inextensibility of hard tooth tissue will trigger severe pain. The results of this study show that patients report that they experience mild pain when having seizures on apparently healthy teeth. The duration of the seizures of the increasingly regular frequency leads to drainage of inflammatory exudate, resulting in the disappearance of pain and a progression towards pulp necrosis. As a result, dental pain is less and less noticeable for patients with more than three attacks a year.

Dental manifestations of sickle-cell anaemia

Various experiments have shown hypomineralizations of enamel and dentin and periapical bone lyses [12,13]. This hypomineralization of hard tooth tissue is a weakness criterion that favours a rapid evolution towards pulpopathy in the event of bacterial attack. In another sense, sickle cell disease is characterized by hemoglobinopathy, which results in a deficiency of red blood cells, the vectors of an element such as blood iron. The correlation between iron deficiency anaemia and high susceptibility to developing carious disease is admitted [14,15]. This investigation is consistent with the data from the literature, showing that of the 65% of patients affected by carious disease, 60% have a deficiency of red blood cells. The presence of sickle cell anaemia in the dental pulp during seizures and pulp calcifications appears to be the cause of significant pulp manifestations compared to dentinal and periapical damage [9,12,13]. The impact of the disease on periapical lesions is still unclear but pulp necrosis is a bacterial source capable of initiating apical periodontitis. The results show that all periapical manifestations are associated with decayed teeth [6].

Patient management

There is no current evidence that dental pain outside of the vaso-occlusive crisis is a potential prodrome of dental pain. It seems that, on the other hand, sickle-cell anaemia promotes the onset of dental pain, the weakening of hard tissues and ultimately tooth pathologies. From presumption to certainty, a correlation has been established to date between sickle cell anaemia and pulp necrosis [10]. The management plan for treatment in the Ivorian context consists of:

(1) identifying and removing the triggering factors for seizures
(2) relieving pain with non-steroidal anti-inflammatory drugs (NSAIDs)
(3) assessing the need for blood transfusion (systematic when the hemoglobin level is less than 6 g/dl)
(4) restoring the rheological properties of red blood cells with vasodilators.

The contribution of an odontological examination from the early stages of management (Phase 1 and Phase 2) could make it possible to treat initial lesions and instruct patients in oral hygiene advice in order to prevent complications of these initial lesions. Systematic integration of an oral examination into the hematological control check-up is one of the key features of this framework. Systematic integration of an oral examination into the hematological check-up is one of the ways.

Conclusion

This study shows that sickle cell patients have dental manifestations with type of dental pain during vaso-occlusive

and hemolytic seizures. Dental pain without carious lesions is characterized by symptomatic irreversible pulpopathy that tends to disappear with the recurrence of sickle cell attacks and progress to pulp necrosis. Subjects with low levels of red blood cells appear to be more likely to develop carious disease. The interest of this investigation is to propose a multidisciplinary treatment of patients suffering from SSFA2 homozygous sickle-cell anaemia by integrating a systematic oral examination, from the diagnosis of the disease to the different phases of surveillance.

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