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Case series

Health-related quality of life in patients with noma (cancrum oris): Case series

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ABSTRACT

Introduction and importance: Cancrum oris (noma) is a destructive disease of orofacial tissues and adjacent structures that progress rapidly, causing functional and cosmetic problems to the patient and ultimately affecting their health-related quality of life. This case series focuses on the impact of the cancrum oris on the health-related quality of life of patients which has been underreported in literature.

Case presentation: Herein, we present 3 cases (1 pediatric and 2 adult patients) of cancrum oris. One adult male succumbed to the disease while the remaining two are undergoing treatment. We used the modified versions of the Early Childhood Oral Health Impact Scale (ECOHIS) and the University of Washington-Quality of Life (UWQoL) questionnaire for pediatric and adult patients respectively.

Clinical discussion: The sequela of cancrum oris affects negatively the quality of life of the patients even after definitive management. Despite this, there is no validated questionnaire to assess the quality of life. The modified versions of the ECOHIS and the UWQoL questionnaires that were used in this case report can be used effectively to assess the HRQoL of the patients.

Conclusion: Cancrum oris is a neglected disease that affects negatively the health-related quality of life of patients who suffer from it, despite medical intervention.

1. Introduction

Cancrum oris (noma) is a destructive disease of orofacial tissues and adjacent structures that progresses rapidly [1]. It primarily affects children and rarely adults [2]. The main risk factors for the disease are poverty and impaired immunity of the body [3]. Apart from its high mortality rate of up to 90 % in untreated cases [4], patients with cancrum oris usually suffer from functional and cosmetic problems [5] which severely affect their health-related quality of life (HRQoL). Health-related quality of life (HRQoL) has been defined as the functional and psychosocial outcome of an ailment and its subsequent management upon the patient [6]. The majority of published literature on cancrum oris concentrates on the epidemiology, etiopathogenesis, clinical presentation, and management of the disease [1,2,5,7]. However, the impact of this devastating disease on HRQoL is underreported globally. Therefore, clinicians have a poor understanding of the disease from the patient's point of view. In light of this, this article reports a series of cases of cancrum oris (noma) focusing on the impact of the disease on the HRQoL of patients. These reported cases are of the patients seen in the department of oral and maxillofacial surgery at Muhimbili National Hospital, a national referral hospital that is located in Dar es Salaam, Tanzania. This work has been reported in line with the SCARE criteria [8].

2. Case 1

A 2-year-old boy old boy, whose father is a civil servant, presented to us with difficulty in opening his mouth in 2018. His condition was secondary to a necrotic ulcer that had developed around his perioral region. The clinical, laboratory, and radiological investigation pointed out the diagnosis of cancrum oris. The sequela of the disease led to massive fibrosis around the oral cavity, due to which he could barely open his mouth, leading to difficulty feeding. In the initial days of admission, a nasogastric tube (NGT) was inserted for feeding purposes. However, a gastric feeding tube (GFT) was eventually placed since the patient kept on pulling out the NGT.

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In 4 years, a series of surgical procedures have been performed on him. The aim has been the management of his condition and subsequent quality of life. These include GFT placement, commissuroplasty, tracheostomy, and skin grafting around the perioral region. Despite the efforts of the surgical team, the patient, who is currently 6 years old, has minimum improvement in general health and stunted growth.

Concerning the quality of life, a modified Early Childhood Oral Health Impact Scale (ECOHIS) was used to assess the quality of life of the patient. The scale has two main sections: the Child Impact Section (9 items) and the Family Impact Section (4 items), and the response options are coded as 0 = never;1 = hardly ever; 2 = occasionally;3 = often;4 = very often. [9] The guardian of the child responded to the questionnaire, and the score for Child Impact was 29 out of 36, and for Family Impact, it was 15 out of 16 (Table 1).

3. Case 2

A 39-year-old male patient, working as a small-scale fisherman, presented to us with a gaping wound on the left cheek for about two months in 2017. His condition was preceded by a swelling that ulcerated and became necrotic (Fig. 1). Diagnosis of cancrum oris was reached after clinical and radiological investigations. Laboratory investigation revealed he was HIV positive, malnourished, had deranged serum electrolytes, and was anemic.

The management plan included optimizing the patient before definitive surgical management. Institution of broad-spectrum antibiotics therapy, hydration for correcting electrolyte imbalance, nutritional support, and blood transfusion were done. Moreover, debridement of the necrotic tissues and daily wound care was performed. However, after one week of admission, the patient's condition deteriorated and he succumbed to death.

Table 1
ECOHIS response of the child's parent.

	Response to the questions
Impact on the child	
Child symptoms	
1. How often has your child had PAIN in the teeth, mouth, or jaws	1
Child function	
How often has your child …… because of the disease or its management?	
2. Had difficulty DRINKING hot or cold beverages	4
3. Had difficulty EATING some foods	4
4. Had difficulty PRONOUNCING any words	4
5. Had missed preschool, daycare, or school (ABSENCE)	4
Child psychology	
How often has your child …… because of the disease or its management?	
6. Had trouble SLEEPING	1
7. Been irritable or FRUSTRATED	3
Child self-image and social interaction	
How often has your child …… because of the disease or its management?	
8. Avoided SMILING or LAUGHING	4
9. Avoided TALKING	4
Impact on family	
Parental distress	
How often have you or another family member …… because of the disease or its management?	
10. Been WORRIED/UPSET	4
11. Felt GUILTY	3
Family function	
How often have you or another family member …… because of the disease or its management?	
12. Taken time off from WORK	4
13. How often has your child's oral health problems or its management had a financial impact on your family FINANCIAL	4
Sub Score for impact on the child	29
Sub Score for impact on family	15
Total Score	44

Fig. 1. Left cheek deep necrotizing ulcer with raised margins, with necrotic base and exposed underlying tissues.

During admission, the quality of life of the patient was assessed using a modified version of the University of Washington-Quality of Life questionnaire [6]. The total score for the patient was 533.2 of the possible 1300 points (Table 2).

4. Case 3

In 2022, a 44- year old male was referred to our department with a gaping defect in his right cheek accompanied by exposure of the bone in his lower jaw (Fig. 2). The patient who is a peasant, is a known case of HIV/AIDS for six years but defaulted on the treatment two years ago. After clinical and radiological examination, the diagnosis of cancrum oris was reached. Laboratory investigation pointed out that he was anemic, was malnourished, and his CD 4 counts were 59 cells/μL.

Table 2
Scores for each domain of modified UW-QoL in patients with NOMA before and after treatment.

Domain name	Cases			
	Case 2		Case 3	
	Pre-operative	Post-operative (1 month)	Pre-operative	Post-operative (1 month)
1. Pain	50	–	50	25
2. Appearance	25	–	25	50
3. Activity	25	–	25	50
4. Recreation	75	–	75	75
5. Mood	25	–	25	50
6. Swallowing	66.6	–	66.6	66.6
7. Speech	66.6	–	100	100
8. Taste	100	–	100	100
9. Saliva (drooling)	0	–	33.3	66.6
10. Anxiety	0	–	33.3	100
11. Chewing	50	–	50	100
12. Mouth opening	50	–	50	50
13. Financial difficulties	0	–	0	0
Total score	533.2	–	633.2	833.2

Key: For the item number one to five: scoring was as follows A = 100, B = 75, C = 50, D = 25, and E = 0. The item number six to ten were scored as A = 100, B = 66.6, C = 33.3, and D = 0. While Item eleven to thirteen were scored as A = 100, B = 50, and C = 0. Zero represented the worst score while a score of 100 represented the best score.

Fig. 2. A deep ulcer on the lower face with raised margins, with necrotic base, and exposed bone.

The management plan included optimizing the patient before definitive surgical management. A broad-spectrum antibiotics therapy, antiretroviral therapy, and nutritional support were immediately initiated. Debridement of the necrotic tissues and daily wound care was performed. After 6 weeks of optimizations, the patient was planned for surgical closure of the defect using local-regional flaps. Unfortunately, apparent wound dehiscence occurred after 5 days of surgery, which was managed conservatively with daily wound care. The patient was discharged and is being closely followed up with fairly acceptable results.

The disease has affected his quality of life negatively. The modified UW-QoL questionnaire was used to assess his HRQoL both on admission and a month post-operatively. The total pre-operative score was 633.2 and the post-operative score was 833.2 out of the possible 1300 points (Table 2). And after the treatment, he still has to attend the hospital on weekly basis, thus missing his work hours.

5. Discussion

Cancrum oris (noma) is one of the oldest diseases to affect mankind [10]. Its risk factors, pathobiology, clinical presentation, and management have been extensively documented in literature from around the globe [1,2,5,7]. However, there are limited studies that report on the HRQoL of patients suffering from this disease.

In the literature, the documented risk factors for noma include poor living standards, extreme poverty, poor oral hygiene, severe malnutrition, and HIV infection [11,12]. Similarly, in this case series, the predisposing factor for cancrum oris in both adult patients was HIV infection, whereas poor nutritional status was a risk factor in the pediatric patient.

There are numerous sequelae of cancrum oris such as difficulty feeding, opening the mouth, speech problems, and drooling of saliva [13]. These sequelae were evident in all three patients in this case series leading to a lower HRQoL in them and subsequently pushing them into seeking medical care. The management of cancrum oris requires multistage surgical treatments that can take years to complete and a longer duration of hospitalization [13]. Similarly, in this report, the pediatric patient has been operated upon several times and has been under treatment for 4 years, while one of the adult patients was hospitalized for nearly 3 months, and is being followed-up bi-monthly.

The longer hospitalization time and cost of management further lead to extreme poverty. To cope with the costs of the disease, patients or caretakers end up spending little savings they had and often sell their properties. Moreover, the patient or their caretakers tend to take time off from their business which in turn impacts their income negatively [12]. This was evident in our cases as well. In 1st case, the caretaker of the child was worried about the finances, and similar concerns were in both adult patients.

Since there are limited studies that look into the HRQoL of the patient with cancrum oris, there is no acceptable validated, and reliable tool that can be used in these patients. In the pediatric patient, we modified and adopted the Early Childhood Oral Health Impact Scale [9]. Whereas, in adult patients, we used a modified version of the University of Washington-Quality of Life questionnaire [6]. Both questionnaires, though not designed for cancrum oris, captured the required information regarding the quality of life in these patients.

The mortality rate of cancrum oris is not well known, though it is estimated to be around 10–20 % if early intervention is initiated [10]. One of the cases in this series succumbed to the disease. Similarly, Pedro et al. [4] reported a case of cancrum oris in a South African adult patient, who succumbed to illness as well.

This case series serves as a reminder to health professionals that the management of cancrum oris should address the mental and social well-being of the patients apart from the esthetic and functional challenges posed by the disease [14]. The scientific community should as well consider developing a reliable questionnaire that would address the health-related quality of life in patients with cancrum oris, despite being a rare disease.

6. Conclusion

Cancrum oris is a neglected disease that affects negatively the health-related quality of life of patients who suffer from it, despite medical intervention.

Consent

Written informed consent was obtained from the respective parents/caretakers of each patient for the publication of the case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethics approval and consent to participate

The study was approved by the Institution Review Board of the Muhimbili University of Health and Allied Sciences (MU/PGS/SAEC/Vol.X).

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Karpal Singh Sohal: Conceptualization, Methodology, Validation,

Resources, Data curation, Writing – original draft, Writing – review & editing. **Arnold A. Mtenga:** Conceptualization, Resources, Writing – review & editing, Supervision. **Dorah Kiwale:** Investigation, Resources, Data curation, Writing – review & editing.

Declaration of competing interest

N/A.

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