

CASE REPORT

A noma case report: A warning message from Northern Uganda

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Abstract

Noma, or *Cancrum oris*, is a severe and rapidly progressing gangrenous infection that primarily affects the face. It is most commonly observed in children living in impoverished conditions, especially in sub-Saharan Africa. Rapid diagnosis and early management are crucial to prevent devastating consequences, such as functional limitations and serious psychological repercussions. Herein, we present a case of an 8-month-old child affected by noma, whose positive outcome is attributed to the prompt recognition by healthcare personnel. In our patient, the condition was likely related to malnutrition and the preceding extraction of a deciduous tooth reported by the mother and probably associated with a traditional Ugandan practice called *Ebiino*. This is the second case reported in Uganda, and given the limited healthcare access in most of the country, coupled with the high prevalence of poverty and other predisposing factors, it becomes evident that the incidence of noma is underestimated. Noma, as a neglected disease, requires greater awareness within communities and among healthcare professionals. A collective effort is needed to significantly reduce risk factors and promote prevention of this life-threatening disease.

KEYWORDS

Cancrum oris, *Ebiino*, gangrenous infection, Noma, Uganda

INTRODUCTION

Noma, also known as *Cancrum oris*, is a polymicrobial, treatable infection of the oral cavity with a gangrenous evolution towards rapid and extensive disruption of orofacial structures [1]. If left untreated, the consequences can be devastating, resulting in a 90% mortality rate within weeks after onset [2]. Therefore, prompt diagnosis and rapid management are crucial to prevent such outcomes and the severe sequelae occurring in noma survivors. Indeed, the rapid spread of the infection can result in facial disfigurements with functional limitations, such as feeding and speech impairment, as well as leading to serious psychological repercussions [3].

Often referred to as 'the face of poverty' [4], noma is predominantly found in children living in low-income countries in Africa and Asia [5]. In fact, reported risk factors include chronic malnutrition, poor hygiene, young age, and previous infections, such as malaria [3]. The United Nations Human Rights Council Advisory Committee has defined noma as a

violation of human rights, serving as a stark reminder of the disparities in the global distribution of resources [6]. Although a huge number of publications originates from Western Africa, the prevalence on the rest of the continent—and namely in Uganda—is largely unknown, as this disease remains unrecognised, under-diagnosed and under-reported. Noma has recently been added to the World Health Organization (WHO) list of Neglected Tropical Diseases (NTDs), acknowledging it as one of the most overlooked health challenges [7].

Herein, we report a case of an infant affected by noma, who was admitted to Dr. Ambrosoli Memorial Hospital, a private not-for profit 350-beds hospital in Kalongo, Northern Uganda, with the aim of shedding light on a neglected disease, not well reported or described in this area, which requires high clinical awareness and early recognition.

CASE REPORT

An 8-month-old girl from Agago district, Northern Uganda, was admitted to the surgical ward of Dr. Ambrosoli

Memorial Hospital in Kalongo due to a necrotic wound affecting the left mandibular area. The condition started 6 days earlier as mild inflammation of the lower left gum, where the extraction of a deciduous teeth had been performed 5 days before the onset of symptoms. Past medical record showed no other notable events. Parents did not report vomiting or breastfeeding impairment. Information regarding prior vaccinations were lacking.

On physical examination, her anthropometric data (weight 5.5 kg, height 66 cm, MUAC 11.3 cm and a Z-score -3 SD) were indicative of severe acute malnutrition according to the WHO definition. She presented with fever and mild anaemia (haemoglobin 8.8 g/L) and she tested negative for HIV. A well-demarcated ulcer, about 8×10 cm wide, involving the subcutaneous tissues and covered by an eschar was detected in the left mandibular area, extending to the left cheek and the submandibular region (see Figure 1a). No other pathological findings were reported on physical examination. The diagnosis of noma was established based on the clinical features of the wound, the patient's medical history and comorbidities.

Extensive surgical debridement was performed on the second and fourth days from admission. Necrotic tissue was aseptically removed up to the lateral side of the left cervical region, exposing the lower jaw and the base of the tongue (b).

Dressing and disinfection with iodine and vinegar were performed daily and, as the wound improved, every other day. Antibiotics were initiated upon admission, consisting of intravenous cloxacillin 250 mg every 6 h for 10 days and intravenous metronidazole 7.5 mg/kg every 8 h for 15 days. She was fed a milk-based formula through nasogastric tube while undergoing close nutritional monitoring. Adjunctive therapy with folic acid was administered, along with paracetamol for pain control.

The general condition and vital signs remained stable throughout hospitalisation and the initial low-grade fever subsided after a few days. Throughout the treatment, the occurrence of a small cervical abscess near the wound, associated with fever, was reported, but healed spontaneously. No major complications were reported.

As the general conditions improved, oral supplementation with ready-to-use therapeutic food was initiated. The patient was discharged after 51 days of hospitalisation with the recommendation to continue wound dressing at the local Healthcare Centre and to attend monthly follow-up appointments at the surgical ward of Kalongo Hospital. At the 2-month follow-up (see Figure 1d), no major sequelae were reported with the exception of temporomandibular joint stiffness on the affected side and mild impairment in chewing.

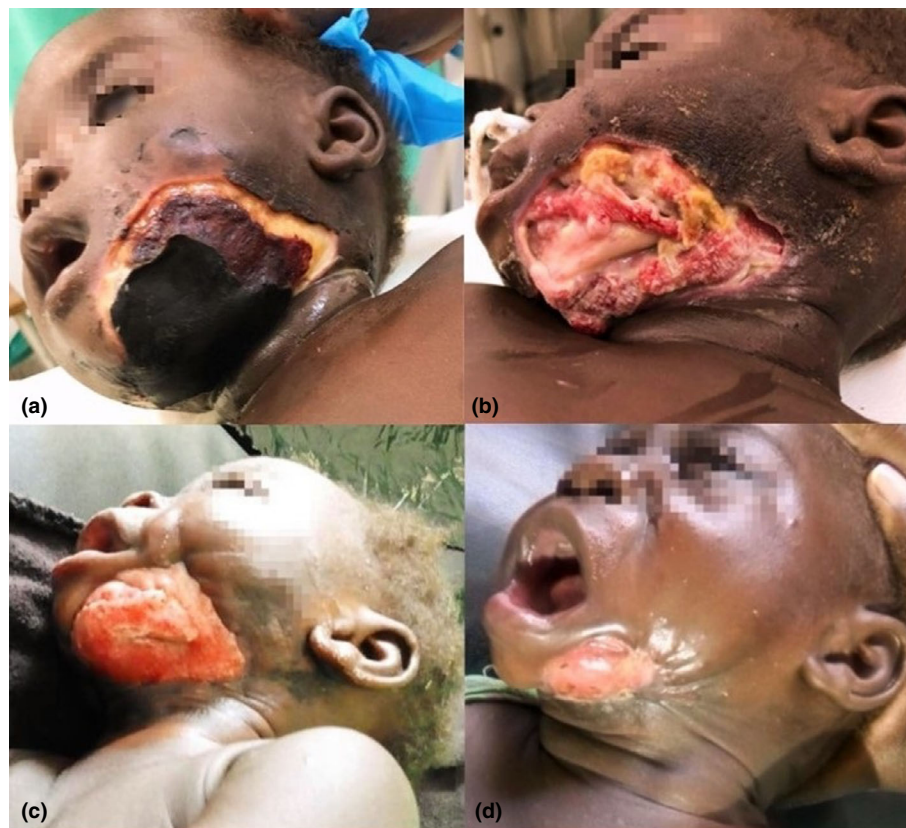


FIGURE 1 The 8-month-old girl admitted to the surgical ward due to a well-demarcated ulcer of the left mandibular and submandibular region, involving the subcutaneous tissues and covered by an eschar (a). The necrotic tissue was removed up to the lateral side of the left cervical region, exposing the lower jaw and the base of the tongue (b). Dressing was performed daily after the surgical debridement, leading to the improvement of the wound as showed in (c) and (d), respectively after 3 weeks and 2 months.

DISCUSSION

Noma has been described as a severe and rapidly progressing infection of the oral cavity that leads to orofacial gangrene and severe deformity. To the best of our knowledge, in this paper we present the second case of noma reported in Uganda. The patient, an 8-month-old child, was diagnosed and treated in the setting of a private not-for profit hospital in Agago, a rural district in Northern Uganda. In this case, as in many others, the disease developed in a resource-limited setting with the risk of being unrecognised or misdiagnosed because of patient's limited access to healthcare facilities or lack of recognition by healthcare professionals.

In the past, noma was common in North America and Europe but gradually disappeared with economic and public health developments [8]. During the decade 2010–2019, the cases reported were from 23 different countries, the majority of which are low- and middle-income countries belonging to the African and Asian continents [9]. In Africa, a disproportionate number of publications originates from Western countries (Nigeria, Niger, Burkina Faso, Mali and Togo), reflecting the work of non-governmental organisations in these areas. The prevalence on the rest of the African continent remains largely unknown, but this does not imply that noma is non-existent there [9]. In 1998, the WHO estimated that 140,000 new cases occur each year globally, with a prevalence of 770,000 people affected [10]. However, these data do not provide an accurate estimation of global noma distribution since only those patients who are able to reach a hospital are taken into account.

The pathogenic mechanism of noma starts in the mouth as bacteria-induced necrotising gingivitis. Several organisms have been identified in the oral flora of affected patients, such as *Fusobacterium necrophorum*, *Prevotella intermedia*, alpha-hemolytic streptococci, actinomyces and peptostreptococci [11]. Nevertheless, none of them have been consistently present, suggesting noma could be caused by a change in the equilibrium of commensal bacteria due to a deterioration in the host defences. The infection begins with inflammation of the gums, which then leads to ulceration and the rapid destruction of the cheek, jaw, lips, nose and/or the eye due to necrotising stomatitis [3]. The WHO has classified noma into stages: Stage 0—simple gingivitis, Stage 1—acute necrotising gingivitis, Stage 2—oedema, Stage 3—gangrene, Stage 4—scarring and Stage 5—sequelae [2]. The first four stages represent the acute phase of the disease, lasting only a few weeks. The sequelae include difficulties in eating, seeing and breathing due to deformity. Trismus (a restriction in mouth opening) is one of the most disabling sequelae [12].

Any child, especially between 2 and 6 years old, with an ulcer in the perioral region and other warning signs such as malnutrition, poor hygiene, recent illnesses like measles, persistent diarrhoea, or malaria should be considered potentially having noma [13]. The lack of access to basic healthcare and the prevalence of traditional practices in rural areas are other potential risk factors for the development of noma [8].

In this regard, *Ebiino* ('false teeth' in Acholi, Northern Ugandan local language) is a traditional practice consisting of the extraction of deciduous canine tooth buds in children. It is based on the belief that the gingival swelling that occurs during their eruption is responsible for common childhood illnesses such as fever, cough and diarrhoea. *Ebiino* can be considered a type of oral mutilation performed by traditional healers, using unsterile instruments and fingernails, leading to complications such as septicaemia [14]. This practice is relatively common in African countries. In Uganda it was first reported among the Acholi people in Northern region [15]. Complications arising from the *Ebiino* procedure have been shown to be the eighth most frequent cause of admission to the paediatric ward in a Northern Ugandan hospital [16] and among these complications, noma has also been described [14]. Besides, the only reported case of noma in Uganda is related to this practice [14]. Even in the case of our patient, an extraction of deciduous teeth had preceded the appearance of the ulcer. Although it was not possible to trace the exact details of the performed procedure, it is reasonable to surmise that it was very similar to that described in the literature and related to *Ebiino*. Considering the low healthcare access, especially in Northern and Eastern Uganda [17], as well as the prevalence of conditions such as malnutrition and these traditional practices, it seems clear that the incidence and prevalence of noma in this country are underestimated.

The rapid progression of noma makes its early identification mandatory to decrease mortality and morbidity. The treatment of early reversible stages of disease consists of administration of broad-spectrum antibiotics (amoxicillin and metronidazole), oral hygiene, wound cleaning, debridement/dressing and nutritional support (high protein). Appropriate reconstructive surgery and physiotherapy are needed in survivors with severe sequelae [12].

The positive outcome in our case is attributed to the surgical team's prompt recognition and proper management, underscoring the importance of training healthcare personnel about this disease. Moreover, healthcare professionals must raise awareness within communities about noma and its risk factors, promoting early recognition of noma signs and discouraging harmful traditional practices, such as *Ebiino* [14].

In December 2023, noma was included in the WHO list of NTDs, following a recommendation of the Strategic and Technical Advisory Group for Neglected Tropical Diseases [7]. The request was supported by a detailed dossier demonstrating noma fulfilment of the WHO inclusion criteria, along with letters of endorsement from 31 countries [7]. The hope is that the inclusion of noma in the WHO's list will prompt allocation of new investments and resources, stimulate research and implementation of prevention and control programs.

Despite this decision represents a crucial step to amplify global awareness on the topic and catalyse political commitment, there is still a long way to go. Indeed, reducing noma cases will require collective efforts involving governments,

humanitarian organisations, healthcare professionals and communities themselves to significantly decrease risk factors, such as malnutrition. Additionally, promoting prevention through, for example, vaccination programs for diseases like measles, which have been linked to noma, will be essential.

CONCLUSION

Noma remains a significantly neglected disease in both research and the global health community. Despite similar socioeconomic, nutritional and healthcare system limitations, there is a disproportionately higher number of reported cases of noma in Western Africa compared to the rest of the continent, including Uganda. The prevalence in this area is still unknown, emphasising the importance of raising awareness among healthcare professionals for its prompt recognition. Furthermore, the implementation of possible prevention programs to mitigate risk factors could compensate for limited healthcare access and discourage the use of harmful traditional practices, such as *Ebiino*. The evident functional consequences experienced by noma survivors, encompassing pain, disability and social stigma, underscore the necessity for the proper management of these cases and their referral to specialised hospital, especially for surgical intervention.

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