

# Noma: Life Cycle of a Devastating Sore — Case Report and Literature Review

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## ABSTRACT

Noma (cancrum oris) is an orofacial gangrene, which during its fulminating course causes progressive and mutilating destruction of the infected tissues. The disease occurs mainly in children with malnutrition, poor oral hygiene and debilitating concurrent illness. Noma is well documented in the literature, but because most patients do not report to a doctor until the disease is at an advanced stage, its onset and progression remain a mystery. This case report, with a survey of recent relevant literature, highlights the different stages in the development of tissue necrosis, including onset and progression, with an emphasis on the need for early diagnosis and prompt treatment.

**MeSH Key Words:** child; noma/complications; noma/etiology

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Cancrum oris or noma (from the Greek *nomein*, “to devour”)<sup>1</sup> is a “gangrenous affection of the mouth, especially attacking children in whom the constitution is altered by bad hygiene and serious illness especially from the eruptive fevers, beginning as an ulcer of the mucous membrane with edema of the face extending from within out, rapidly destroying the soft tissues and the bone and almost always quickly fatal.”<sup>2</sup>

The epidemiology of noma has not changed much over the years, except that there has been a reduction in the mortality rate from 90% to about 8% to 10%, mainly because of modern antibiotics.<sup>3,4</sup> Noma is considered to represent the “face of poverty” because factors connected with poverty, such as chronic malnutrition, poor oral hygiene, poor environmental sanitation, exposure to animal and human fecal material, and exposure to viral and bacterial infections,<sup>1,4,5</sup> contribute to disease progression. The World Health Organization has reported alarming increases in the incidence of noma over the past 10 years,<sup>1</sup> yet little is known about the initial stages and progression of the disease because

most patients with early-stage noma are examined by primary health care workers<sup>1</sup> who do not refer them to city hospitals until later in the course of the disease. The disease has a rapid clinical course, and as a result many patients have advanced noma by the time they are brought to a hospital. In some communities, noma is regarded as a curse or a spell cast on the family, so those affected are ignored rather than being brought to medical attention in the initial stages.<sup>1,6</sup> This report describes a patient in whom all stages of the disease were observed. The current literature is briefly reviewed with particular reference to the causes, diagnosis and treatment of noma.

## Case Report

A 9-year-old boy presented with a painful swelling of 4 days' duration on the right side of his face. He had noticed an ulcer at the corner of his mouth 1 week earlier, after which the swelling appeared and progressed rapidly. He also reported discharge of pus from the corner of his mouth and difficulty in opening the mouth. He had intermittent fever and gave



**Figure 1:** Day 1. Extraoral photograph shows the swelling and discharge of pus from the corner of the mouth.



**Figure 2:** Day 7. Flare-up of infection, with crater-like erosions.



**Figure 3:** Day 15. Well-demarcated area of necrosis, with bluish black discolouration at the periphery.



**Figure 4:** Day 30. Healing lesion, with no signs of active infection.



**Figure 5:** Day 56. Complete necrosis, with exposure of the underlying bone.

a history of chronic cough since childhood. There was no history of trauma.

The patient was malnourished and weighed only 11.5 kg. His temperature was 39°C at the time of the initial check-up, but all other vital signs were within normal limits. The patient had pallor and clubbing of the fingers.

Extraoral examination revealed a tender 4 × 6 cm swelling on the patient's right cheek. The swelling extended from the corner of the mouth to the preauricular region and from the infraorbital region to the lower border of the mandible. There was a local increase in temperature over the swelling. Mild crusting was observed on the lips and at the corner of the mouth (Fig. 1). The skin over the swelling was smooth, stretched and shiny. No scars were observed. The right submandibular lymph nodes were palpable, tender and mobile.

Intraoral examination revealed a 2 × 1 cm ulcer in the right buccal mucosa, extending from the right corner of the mouth to the premolar region. There was no foul

odour, nor were there any dental or periodontal foci of infection. A complete intraoral examination was not possible because of restricted mouth opening.

The initial presentation was suggestive of cellulitis. Panoramic radiography was performed to check for dental or periodontal foci of infection, but no abnormalities were evident.

Hematologic investigations revealed anemia and an elevated erythrocyte sedimentation rate (Table 1). On the first day, an incision was made under local anesthesia; drainage performed by an intraoral approach yielded very little pus. Oral antibiotics and analgesics were prescribed (Table 2), and the patient was admitted to hospital under the care of a pediatrician for evaluation of chronic cough and anemia. The pediatrician reported that the patient had grade II clubbing of the fingers with bilateral crepitations; chest radiography revealed radiopacity in the right paracardiac region, and a provisional diagnosis of bronchiectasis was made.

**Table 1** Results of investigations

Type of test	Patient's value	Normal range
<b>Laboratory tests<sup>a</sup></b>		
Hemoglobin	3.72 mmol/L	8.1–11.2 mmol/L
Erythrocyte sedimentation rate	130 mm/h	4–10 mm/h
Sweat chloride	> 118 mEq/L	< 60 mEq/L
<b>Microbiology tests</b>		
Culture and sensitivity <sup>b</sup>	Multiple gram-negative anaerobic bacteria, sensitive to vancomycin, cefazolin, ciprofloxacin, amikacin and gentamicin but resistant to penicillin	NA
Fluorescence microscopy for Acid-fast bacilli	Negative	NA
Enzyme-linked immunosorbent assay for HIV	Negative	NA

NA = not applicable

<sup>a</sup>The results of all other laboratory investigations were normal.

<sup>b</sup>Because anaerobic culture media were lacking, a sample was sent on day 4 of admission for culture and sensitivity.

The swelling did not progress further over the next 2 days. After a week (on day 7), the infection flared up (Fig. 2), and 2 weeks later bluish black discoloration developed at the margins of the lesion. At this stage, a definitive diagnosis of noma was made. The pediatrician later requested estimation of sweat chloride levels (Table 1), and cystic fibrosis was diagnosed on day 23.

The complete progression of the disease and adjustments in treatment planning are detailed in Table 2.

## Discussion

Some unusual presentations of noma have been described in the literature. Noma neonatorum is a rare gangrenous form affecting the oronasal tissues in medically compromised newborns at birth or during the first month of life.<sup>7,8</sup> Noma pudendi is another form affecting the anogenital area and causing necrosis of the genitalia.<sup>9</sup>

In 90% of cases, noma develops before the age of 10 years.<sup>10</sup> Infants and young children are less susceptible, because breast milk provides protein and protective antibodies for infants and young children; children become susceptible to the disease after cessation of breastfeeding or if their diet is deficient in proteins, which weakens immunity. The patient described here was a 9-year-old boy whose diet was severely deficient in proteins and vitamins.

Noma is not a primary disease. Predisposing illnesses such as measles, tuberculosis, leukemia and AIDS usually precede its occurrence.<sup>1,2,11</sup> An increasing incidence of noma and noma-like lesions in people with AIDS has also been reported.<sup>12</sup> The patient described here had bronchiectasis, as well as cystic fibrosis, which affects the

pancreas and intestinal tract, causing intestinal obstruction and malabsorption;<sup>13</sup> this situation explains the patient's malnutrition and stunted growth. Malnutrition leads to alteration in cell-mediated immune function and early breakdown of the epithelial tissues; alterations in the oral mucosa facilitate invasion by pathogens.<sup>1</sup> Eating difficulties due to infection further aggravate any existing malnutrition. Stresses on malnourished children living in poor environmental conditions increase the level of circulating cortisol, which initiates a cascade of reactions that impair the immune system and favour the growth of bacteria.<sup>1,5</sup> Therefore, malnutrition can be considered to have been a major predisposing factor for development of noma in this patient.

It is difficult to pinpoint the specific trigger agent in the complex microbiota of a noma lesion. It has been speculated that *Borrelia vincentii* and *Fusobacterium* are prominent bacteria in such lesions.<sup>14,15</sup> Symbiotic relationships between fusiform bacilli and non-hemolytic streptococci and staphylococci have been considered significant factors in the development of noma.<sup>1</sup> Previous studies were hampered by a lack of specimens from early stages of the lesion, confusing microbial taxonomy, lack of animal models for studies and difficulty in culturing microorganisms.<sup>14</sup> Recent reports suggest that besides fusiform bacilli and spirochetes, other anaerobic bacteria are present in a relatively high proportion of noma lesions.<sup>15</sup> *Fusobacterium necrophorum* is considered a key component; this organism produces dermatotoxins, which could explain the rapid progression of the disease.<sup>16</sup> *Fusobacterium necrophorum* is acquired by impoverished children through fecal contamination of water, which

**Table 2** Chronology of events leading to tissue destruction and necrosis<sup>a</sup>

Time after admission	Symptoms and signs	Treatment	Notes
Day 1	Tenderness, swelling of 4 days' duration, pus discharge, fever ( <b>Fig. 1</b> )	Incision and drainage, oral antibiotics <sup>b</sup>	
Days 2 and 3	Tenderness, crusting of lips, foul odour	Oral antibiotics, <sup>b</sup> nutritional supplements	No improvement
Day 7	Flare-up of infection ( <b>Fig. 2</b> )	Parenteral antibiotics, <sup>c</sup> transfusion of 2 units of blood, nutritional supplements	Remission of infection and inflammation
Day 9 <sup>d</sup>	Formation and induration of granulation tissue	Parenteral antibiotics <sup>e</sup>	Healing
Day 12	Peripheral signs of necrosis, bluish black discolouration, crusting of lips	Parenteral antibiotics, <sup>e</sup> oral antibiotics <sup>f</sup>	Progressive necrosis
Day 15	Well-demarcated blackish area ( <b>Fig. 3</b> )	Parenteral antibiotics, <sup>e</sup> oral antibiotics <sup>f</sup>	Established gangrene; diagnosis of noma confirmed
Day 28	Wound dried, proliferation of epithelial tissue at margins	Oral antibiotics, <sup>f</sup> diet chart, parenteral antibiotics <sup>e</sup>	Healing lesion
Day 30	Healing lesion, no signs of active infection ( <b>Fig. 4</b> )	Oral antibiotics <sup>f</sup>	Patient discharged, with recall after 5 days (but patient missed recall appointment)
Day 45	Soft-tissue breakdown over lesion, with bleeding spots	Parenteral antibiotics, <sup>e</sup> oral antibiotics <sup>f</sup>	Reactivation of lesion; readmission
Day 49	Orocutaneous fistula developed	Parenteral antibiotics, <sup>e</sup> oral antibiotics <sup>f</sup>	No response to treatment
Day 56	Complete necrosis, with exposure of underlying bone ( <b>Fig. 5</b> )	Patient's family insisted on discharge because of various constraints	Patient discharged against medical advice, lost to follow-up

<sup>a</sup>Local wound care: paraffin gauze dressing soaked with Betadine, Eusol and hydrogen peroxide throughout treatment.

<sup>b</sup>Amoxicillin 250 mg three times daily, metronidazole 200 mg three times daily, paracetamol 1/3 tablet (as required), tablet of serrapeptidase, 5 mg twice daily, Betadine mouth gargles, oral hygiene instructions.

<sup>c</sup>Intravenous administration of crystalline penicillin 10,00,000 IU every 6 hours.

<sup>d</sup>Change in antibiotics after results of culture and sensitivity testing.

<sup>e</sup>Intravenous administration of vancomycin 125 mg every 6 hours (diluted with normal saline and given over 1 hour) and gentamycin 30 mg every 12 hours.

<sup>f</sup>Ciprofloxacin 125 mg (1/4 tablet), fluconazole tablet, 50 mg twice daily, erythromycin 125 mg three times daily (1/4 tablet).

occurs when residential facilities are shared with animals and sanitation is very poor. *Prevotella intermedia* has the ability to break down lipid structures, which contributes to tissue destruction. It also produces proteolytic enzymes capable of breaking down immunoglobulin G, which impedes elimination of microorganisms.<sup>1</sup> Some reports have suggested that these microorganisms are resistant to penicillin,<sup>15</sup> as was observed in the patient described here (**Table 1**), which emphasizes the need for culture and sensitivity tests before administration of antibiotics. Also, the disease has a multifactorial etiology, so additional studies are warranted to elucidate its exact microbiology.<sup>17</sup>

On initial presentation, the condition of the patient described here mimicked cellulitis. Therefore, incision

and drainage were performed and oral antibiotics were prescribed (**Table 2**). The flare-up of the infection on the seventh day (**Fig. 2**) was initially attributed to resistance of the microorganisms to the prescribed antibiotic (amoxicillin) and poor systemic health. The diagnosis of cystic fibrosis on day 23 further explained the initial lack of response to therapy (because of malabsorption of oral antibiotics). The results of culture and sensitivity testing were unavailable during the first week of treatment (**Table 1**). Thus, although the patient initially presented with signs and symptoms consistent with noma, the disease was not diagnosed until much later.

After the culture reports became available, aggressive antibiotic therapy was instituted (**Table 2**) to halt the

spread of the oral infection and to treat the lung infection. Parenteral antibiotics, including vancomycin and gentamicin, were started, and the drug dosages were adjusted according to the patient's age to prevent toxic effects. In addition, 2 units of blood were transfused. After the blood transfusion, the patient's general condition improved, and he also responded well to the parenteral antibiotic therapy. Oral ciprofloxacin and fluconazole were also prescribed (Table 2), and the patient was advised to consume plenty of water and other fluids to prevent dehydration. Supplemental parenteral fluid replacement was also provided to maintain electrolyte balance.

Because oral feeding was causing discomfort, nasogastric intubation was performed for administering food, protein and vitamin supplements, and the patient gained 1.5 kg over 1 month of treatment at the hospital. Paraffin gauze dressings soaked with Eusol (Edinburgh universal solution of lime), Betadine and hydrogen peroxide were applied to the wound, and the dressings were changed frequently to remove sloughed tissue and to prevent local wound infection.

The lesion started to heal, but bluish black discolouration developed at the periphery (Fig. 3), a characteristic feature of noma.<sup>3,18</sup> Intervention at this stage with parenteral antibiotics, nutritional supplements and rehydration halted the disease progression (Fig. 4). However, the parents insisted on discharge because of financial constraints, and the patient was discharged against medical advice. At the time of discharge, there were no active signs of infection and the lesion was healing. The patient was advised to continue the antibiotics and dietary supplements (Table 2), and to return for review after 5 days.

The patient failed to return for the scheduled review but reported 2 weeks later with reactivation of the lesion. It was learned that because of the family's financial constraints he had been unable to take the prescribed medications. He was admitted to hospital, and local measures and systemic medications were reinstated (Table 2). However, there was no response to treatment, and an orocutaneous fistula developed. This fistula later progressed, leading to complete necrosis with exposure of the underlying bone, typical of noma (Fig. 5). Again the parents insisted on discharge against medical advice, and the patient was lost to follow-up.

It has been said that "there is nothing like *noma*,"<sup>2</sup> a statement that certainly holds true for the advanced stages of the disease, when it is easy to diagnose. In the case reported here, however, the initial stages of the disease mimicked soft-tissue cellulitis (Fig. 1). Only when bluish black discolouration developed was the diagnosis of noma considered. The differential diagnosis must include mucocutaneous leishmaniasis, lupus erythematosus, leprosy, agranulocytic ulcerations, physical trauma (including burns), syphilis, oral cancer and yaws (an

infectious tropical disease caused by the bacterium *Treponema pertenuae*).<sup>1,2,19,20</sup>

In this patient local debridement of necrotic tissue was performed under local anesthesia throughout the course of treatment, as his lung infection prevented the use of general anesthesia. Usually in such cases, maxillofacial plastic surgeons repair the defect once the infection subsides. In this case, this course of action was not possible because the patient's parents insisted on early discharge against medical advice, the family had financial constraints, and persistent lung infection prevented any surgical procedure under general anesthesia.

Noma can be treated successfully but it leaves behind an ugly scar because of complications. The orofacial tissues grow rapidly during childhood, but the normal developmental process is affected in children who have had noma. The disease can cause premature loss of deciduous teeth, damage to the permanent tooth buds, sequestration of the jaws, trismus, and bony or fibrous ankylosis of the temporomandibular joint.<sup>2,21</sup> The resultant facial asymmetry increases with the growth of the child. The phrase "anarchie dentaire" is used to describe such characteristics.<sup>2</sup> Furthermore, infections from the oral cavity can spread to other parts of the body.<sup>22</sup> Other systemic complications such as toxemia, dehydration and bronchopneumonia can occur and may cause death.<sup>18</sup>

## Conclusions

This case report is of interest because clinicians only rarely get a chance to observe and treat noma from onset of the disease. Recent updates on the causes of the disease and nature of its progression, summarized in this report, may guide the clinician in better managing this devastating disease, which is rapidly emerging as a public health issue in some parts of the world. A team approach to management, involving a dentist, a maxillofacial surgeon and a pediatrician, is needed. In view of the severity of the disease, global efforts are needed to eradicate it. ♦

## THE AUTHORS

*Acknowledgement:* The patient was admitted under the care of Dr. Nalini Bhaskaranand, a professor in the department of pediatrics, KMC, Manipal, India.



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