

Case Series

Cancrum Oris among children seen at Paediatric Medical Ward Ahmadu of Bello University Teaching Hospital, Zaria: A Case series

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Abstract

Background: Cancrum oris (Noma) is an opportunistic infection promoted by extreme poverty. It evolves rapidly from a gingival inflammation to grotesque orofacial gangrene. It occurs worldwide but is most common in Sub-Saharan Africa. The peak incidence of this gangrenous stomatitis is at age 1-4 years coinciding with the period of linear growth retardation in deprived children. Noma is a scourge in communities with poor environmental sanitation that results from complex interactions between malnutrition, infections, and compromised immunity. Diseases that commonly preceded Noma include measles, malaria, severe diarrhoea, and necrotizing ulcerative gingivitis. The acute stage responds readily to antibiotic treatment. The sequelae after healing include variable functional and aesthetic impairments that require reconstructive surgery. Noma can be prevented through the promotion of national awareness of the disease, poverty reduction, improved nutrition, promotion of exclusive breastfeeding, optimal prenatal care, and timely immunization against the common childhood diseases. **Case Series:** Four children aged between 2 and 13 years were admitted and managed for Cancrum oris within 3 months (June –August 2022). Two of the patients had measles and one had both measles and varicella infections. One of them received only the BCG vaccine while the remaining 3 did not receive any form of vaccination. All of them were undernourished and from low socioeconomic backgrounds. They all responded to medical treatment and were subsequently referred for reconstructive surgery. The cases are presented to create awareness.

Keywords: *Cancrum oris, Children, Oral hygiene, Under-nutrition, Zaria*

Introduction

The World Health Organization International Statistical Classification of Disease code A69.0 lists necrotizing ulcerative stomatitis, which includes Noma, Cancrum oris, and fusospirochaetal gangrene. Noma is derived from the Greek νομν,^{1,2} which means to graze or to devour. Orofacial Noma is an infectious disease that starts as gingival ulceration and spreads rapidly through the orofacial tissues establishing itself with a well-demarcated perimeter surrounding a blackened necrotic centre.^{1,3,4} The gangrene can involve not only the mandible and maxilla but also the nose and infra-orbital margins. Unlike other infectious processes of the face which mostly expand along cellular spaces of the head

and neck, the Noma lesion spreads through anatomical barriers such as muscles.

Acute Noma is seen predominantly in children aged 1-4 years, although late stages can occur in adolescents and adults.^{4,5,6} Many patients with Noma present with a range of features reflecting pre-existing, debilitating health conditions. They include fever, tachycardia, tachypnea, and anorexia. The medical history generally shows recurrent fever, diarrhoea, and infections with parasites (malaria) and viruses (measles, herpes) in the recent past.^{6,7} The orofacial lesion can occur unilaterally or bilaterally, but it is unilateral in many cases

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Poverty is the key risk factor for Noma in Africa.^{7,8,9} A retrospective study of 173 cases at a hospital in Nigeria showed that 98 percent were from very poor homes with a mean of seven children per family.^{7,8} Chronic malnutrition is a major predisposing factor in all countries reporting Noma.^{2,3}

It is common in environments with unsafe drinking water, scanty sanitation, proximity to neglected livestock, nomadic lifestyle, and a high prevalence of diseases such as measles, malaria, and diarrhea.^{9,10}

The key points of management during the acute phase of noma are prompt admission into hospital, correction of electrolyte imbalance, nutritional rehabilitation, antibiotics, daily dressing of the lesion with gauze soaked in oral antiseptic, and treatment of associated systemic diseases.^{2,3,5}

Physiotherapy should be initiated during the healing phase and continued after surgery to prevent stricture of the mouth resulting from fibrous scarring.^{6,7}

Case 1

A Z, 2 year-old girl presented with left facial swelling and progressive ulceration, progressive weight loss of 3 months, and leg swelling of 1 week before presentation.

The swelling started on the left cheek, close to the angle of the mouth, initially twice the size of a bean seed, and progressively became diffuse to involve the whole of the left cheek over a week. The swelling was painless and firm, with no bleeding or discharge. Swelling subsequently started ulcerating, with associated darkening of the upper gum. There was a gradual loss of tissues from the left eye, the whole of the nose, the upper jaw, and the adjacent left side of the face leaving the mouth wide open. There was also a gradual loss of teeth in the upper jaw over 2 months, leaving only one on the right side at the time of presentation. Oral hygiene was fair as the mother uses Maclean toothpaste for the child daily. There has been difficulty with feeding the child as she was on only a liquid diet mainly tea and pap in the last 2 months before presentation.

Parents subsequently noticed progressive weight loss. No history of chronic cough or contact with adults with chronic cough, and no recurrent or chronic diarrhea. She had a poor vaccination history, as only BCG was given at 3rd week of life. She had a poor nutritional history and was from a low socioeconomic background. No previous history was suggestive of measles or pertussis-like illness in the child.

On examination, she was ill-looking, irritable, wasted, afebrile, mildly pale, and anicteric. She had brownish silky hair and bilateral pitting pedal oedema, no dermatosis or vitamin A deficiency eye changes.

There was extensive loss of tissue over the mid-facial region to the left side, complete loss of external nose, left lower eyelid, whole of the upper lip, and medial aspect of the left cheek. There was also loss of the maxilla and the hard palate with exposure of the turbinates, nasopharynx, and the oropharynx. Only one tooth was seen on the right side of the upper jaw with intact lower dentitions.

Anthropometric measurements are deranged, weight 8kg (<5th centile), Length 82cm(10th centile), MUAC-11cm (severely malnourished).

Systemic examination was essentially normal.

A diagnosis of severe acute malnutrition with complications of extensive Cancrum facialis was made.

The results of the investigation and treatment are shown in the table below.

The child lost oedema within the first week of admission and started gaining weight. Vital signs remained stable throughout admission.

Following extensive discussion between the managing team, parents, and plastic surgery unit on the cost of facial reconstruction for the patient, she was referred to NOMA specialist hospital, Sokoto under Medecins San Frontier Foundation, for further management after 16 days on admission.

Case 2

UM, 13 year old girl was admitted to EPU, ABUTH Zaria following on presentation with a history of weight loss of 4 weeks duration and ulceration of the right side of her face of 3 weeks duration. The weight loss was preceded by profound anorexia noticed 6 weeks prior following measles and varicella-like illness (both occurred as outbreaks in her neighborhood and affected several family members and neighbors). She had a history to suggest undernutrition as her diet was mostly carbohydrates with minimal consumption of food sources containing protein and vitamins.

Three weeks before the presentation, she developed an ulcer on the right side of her face. The ulcer was located on her cheek and was initially about the size of her thumb

(about 2x2cm) however progressive erosion of the right side of her cheek occurred over 2 weeks period with initial exposure of a fleshy material that eventually sloughed off to reveal the bones and teeth of both the upper and lower jaw of the affected side. This persisted till the presentation. The ulcer was preceded by a painful sore noticed on the inner side of her cheek and a diffused shiny swelling of the lower side of her face noticed 4 weeks prior. This spontaneously ruptured with continuous drainage of foul-smelling purulent discharge that persisted till presentation. There was associated drooling of saliva from the right side of her face and difficulty with chewing as food particles were noted to fall off from the ulcerated side of her mouth.

She was given several over-the-counter medications during her illness and had several other concoctions. She was brought to our facility due to the persistence of the ulcer.

She was unvaccinated and from a low socioeconomic background (her father is a painter with a primary level of education and has 2 wives with 10 children).

Examination findings at presentation revealed a chronically ill-looking adolescent girl, cachexic, stunted, moderately pale, anicteric, afebrile, acyanosed with no significant peripheral lymphadenopathy and no pedal oedema. She had brownish silky hair with a flag sign, widespread hypopigmented skin patches, and desquamation of lower limbs. She had xerophthalmia (clouding of the right cornea and conjunctiva xerosis on the left).

Weight was 13kg (< -3 SD for her age)- severely wasted. Height was 117cm (< -3 SD for her age)-severely stunted. She had xerophthalmia (clouding of the right cornea and conjunctival xerosis on the left). She had an ulcer on the right lower part of her face extending from the tragus of her right ear to the philtrum of her upper lip and middle of her chin exposing both the mandible and maxillary bone of the right side. It measured about 4x8cm, the base of the ulcer, the teeth, and gums of the mandible and maxillary bone, the floor is covered with yellowish purulent discharge and the edge is sloppy. There is marked halitosis, oedema, and hyperemia of the surrounding tissues. Systemic examination: essentially normal.

A diagnosis of Cancrum facialis in severe acute malnutrition was made.

Investigation results and treatment are shown in the table. She had gradual improvement with a weight gain of 5 kg over a month and gradual regranulation of the ulcer on the right cheek. She was scheduled for evisceration of the right eye but was discharged on request for her parent to source funds, for the surgery and other care of the patient. She was however unfortunately lost to follow-up.

Case 3

He is S.M., a 2-year-old boy who presented with a 2-week history of facial ulcer and fever of 2 days duration.

His problem started with a measles-like illness, about 2 weeks later, he developed a facial ulcer, which started as a swelling on the left side of the lower eyelid and progressed to involve the nose, cheek, and upper lip. The swelling was not painful and the overlying skin was shiny with no discharge. However, 8 days later, the overlying skin became dark and sloughed off exposing the bony structures of the eye and nostrils. No preceding trauma or insect bite and no swelling in other parts of the body.

He has never been vaccinated and has a poor nutritional history and poor oral hygiene.

The child is the 9th of the nine children of the mother, 8 alive, 3rd child died at 3 years of age from febrile illness. Mother is a 40-year-old with no formal education and not gainfully employed married to a 50-year-old small-scale farmer in a monogamous setting.

On examination, he was acute on chronically ill-looking, febrile, moderately pale, had hypoproteinemic hair changes, was not dehydrated, had no significant peripheral lymphadenopathy, and had no pedal oedema. Weight 5.7kg (< -3 SD for his age)- severely wasted. Length 77cm (< -3 SD for his age)- severely stunted. MUAC 10.5cm (severely malnourished).

Had a facial ulcer that involved the eye, both upper and lower eyelids, the bridge of both nostrils, the upper jaw, and the right cheek. The floor of the ulcer exposed the nasal turbinates and left eyeball and was covered with a purulent exudate. The edge of the ulcer had some sliding and punched edges. The ulcer is irregular in shape measuring 20x 10 cm in its widest diameter. Systemic examination was essentially normal.

Diagnosis of Severe acute malnutrition with complications of Cancrum oris and sepsis was made.

The investigation results and treatment are shown in the table below.

He improved steadily, the ulcer became clean, and was healing well. There was steady weight gain and he was subsequently discharged home after 8 weeks on admission to be followed up in the clinic.

Case 4

A. M, 5 year old boy presented with an offensive wound on the right cheek of 3 weeks duration and a fever of 1 week duration.

The wound was preceded by a 10-day history of swelling on the right cheek. The swelling was firm but later became soft and ruptured 3 weeks before presentation, it then formed a wound estimated to be 1x1 cm and oozing pus-like material. It began to grow wide and deep with eventual communication with the oral cavity within 10 days of rupture. The wound was very offensive and said to have increased to more than 10 times its initial size. One could visualize the teeth from the site of the wound. The child eats normally with the expulsion of food particles through the wound.

No history of chronic diarrhea, no past measles or pertussis. He was never vaccinated. The child had a poor nutritional history and poor oral hygiene. He was weaned off breast milk at eighteen months of age and taken to his grandmother for continued care. While with his grandmother, his meals were mainly carbohydrate-based.

The fever started 1 week before the presentation. It was described as high grade and intermittent, it persisted till presentation.

At the onset of symptoms, grandmother was applying herbal medication to the wound twice daily without any improvement, he was then brought to ABUTH Zaria.

He is the 1st of the mother's 2 children, all alive. The younger sibling is doing well. The mother is a 20-year-old housewife with no formal education and is not gainfully employed. Father is a 30-year-old truck pusher with no formal education. The child is currently under the care of his grandmother who is a 56-year-old widow with no formal education and not gainfully employed. The grandmother lives in the same household with 4 of her male children who take care of her and the patient.

On examination, he was ill-looking with facial disfigurement and a very offensive /necrotic ulcer on the right cheek, febrile, severely pale, anicteric, acyanosed, not dehydrated, has hypoproteinaemic hair changes with widespread hypopigmented macula patches on the skin, purulent right eye discharge, no pedal oedema. Weight 10kg (< -3 SD for his age)- severely wasted. Height 88cm(< -3 SD for his age)- severely stunted. MUAC-12.5cm. OFC 46cm CC- 47cm. OFC/CC<1(abnormal) Had a necrotic/offensive right cheek ulcer that measured 8x 6cm. It extended from the lower border of the mandible to 1 cm below the right lower eyelid vertically and from the angle of the mouth to about 6 cm from the tragus of the right ear horizontally. It had a necrotic base with a sloughing edge.

Cardiovascular system: PR 150bpm, apex beat at 5th left intercostal space, midclavicular line, Heart sounds were S1 and S2 with 3/6 apical systolic murmur. Respiratory system: RR 40cycles/min (tachypnoeic but not dyspnoeic)

Other systems: Essentially normal

Diagnosis: Severe acute malnutrition with complications of Cancrum oris, sepsis, severe anaemia, and micronutrient deficiency

The available results of the investigation and treatment are shown in the table.

He improved steadily, the anaemia was corrected and the ulcer became clean and was healing well. There was steady weight gain and he was subsequently discharged home after 6 weeks of admission to be followed up in the clinic.

Characteristics of the four patients with Cancrum Oris

Subject	Age (years)	Sex	Risk factors	Weight (kilograms)	Height (centimeters)	Investigations	Treatment	Outcome
A Z	2	F	Poor vaccination, under-nutrition, poor socioeconomic background	8kg (<-3SD for age)	82cm (<-3SD for age)	Hb-8g/dl, U/E/Cr- (normal ranges), retroviral screening, HBsAg and HCV are all negative, wound swab m/c/s cultured pseudomonas specie.	IV Ciprofloxacin, gentamicin and metronidazole, tetanus toxoid, nutritional rehabilitation, oral toileting, and wound dressing with EUSOL.	Did well and was referred to NOMA hospital
U M	13	F	Measles, varicella, under-nutrition, poor oral hygiene, not being vaccinated, and poor socioeconomic background	13 (<-3SD for her age)	117 (<-3SD for her age)	WBC -14.9x 10 ⁹ /l, Lymphocytes-57%, granulocytes- 32.5%, Hb- 9g/dl, wound swab m/c/s- cultured E. coli, ESR-140mm/hr	IV Ceftazidime, metronidazole and gentamicin, Tetanus toxoid, nutritional rehabilitation, oral toileting with chlorhexidine, and wound dressing with EUSOL 1 and 2	Done well and discharged home but lost to follow-up
S M	2	M	Poor oral hygiene, not vaccinated, undernutrition, poor socioeconomic background	5 kg(-3SD for his age)	77cm (<-3SD for his age)	WBC 12x10 ⁹ /L, Hb-8.4g/dl, HIV-negative, wound swab m/c/s cultured E. coli	IV ampiclox, gentamicin, metronidazole, zinc sulphate, folic acid, vitamin A, nutritional rehabilitation, oral toileting with chlorhexidine, and wound dressing with EUSOL	Discharged home and being followed up in the clinic
A M	5	M	Poor oral hygiene, not vaccinated, Geographical weaning, undernutrition, low socioeconomic background	10 (-3SD for age)	88 (-3SD for age)	WBC-4.4 x 10 ⁹ /l, granulocytes-42.7%, lymphocytes- 43%, PCV-10%, wound swab- cultured staphylococcus aureus, U/E- hyponatremia, hypokalemia, HIV- negative	IV ampiclox, gentamicin, and metronidazole, blood transfusion with packed cells, nutritional rehabilitation, wound debridement, and dressing with EUSOL	Discharged home and being followed up in the clinic



Figure 1: Cancrum oris for case 1



Figure 2: Cancrum oris for case 2



Figure 3; Cancrum oris for case 3



Figure 4A; Cancrum oris before treatment for case 4



Figure 4B ; Cancrum oris on treatment for case 4

Discussion

Acute Noma is seen predominantly in children aged 1-4 years, although late stages can occur in adolescents and adults.^{4,5,6} Many patients with Noma present with a range of features reflecting pre-existing, debilitating health conditions.

All the patients presented to our hospital within 3 months (June-August). They are from poor socioeconomic backgrounds, presenting with various degrees of undernutrition. Two had measles and one had varicella infection in addition to measles. There was no vaccination in three of them, but one received only the BCG vaccine. Poor oral hygiene was seen in all of them. Poverty is the key risk factor for noma in Africa.^{7,8,9} A retrospective study of 173 cases at a hospital in Nigeria showed that 98 percent were from very poor homes with a mean of seven children per family.^{7,8} Chronic malnutrition is a major predisposing factor in all countries reporting noma.^{2,3,5-7} All our patients came from poor parents with chronic undernutrition. Noma is common in environments with unsafe drinking water, scanty sanitation, proximity to neglected livestock, nomadic lifestyle, and a high prevalence of diseases such as measles, malaria, and diarrhoea.^{8,9} Two of the patients had measles and one had both measles and varicella infections.

The key points of management during the acute phase of noma are prompt admission to hospital, correction of electrolyte imbalance, nutritional rehabilitation, antibiotics, daily dressing of the lesion with gauze soaked in oral antiseptic, and treatment of associated systemic diseases.^{8,10}

While on admission, all the cases had wound debridement and daily dressing with EUSOL. They were commenced on antibiotics (parenteral ampiclox, gentamycin, and metronidazole) and micronutrient supplementation like zinc sulphate, folic acid, vitamin B complex, and vitamin A. Patients were dewormed and also treated for malaria. Nutritional rehabilitation was instituted. They were co-managed with plastic and maxillofacial surgeons.

They improved steadily, and the ulcers became clean and were healing well. There was steady weight gain and they were subsequently discharged home after 6 to 8 weeks on admission to be followed up in the clinic while being worked up for surgery.

Conclusion

Poor oral hygiene, malnutrition, debilitating diseases (measles and varicella), and lack of vaccination have

been identified as predisposing factors to the development of Cancrum oris in our patients. Health education and economic empowerment are still required to curb the menace of this devastating disease.

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