

The real face of “face of poverty”: an insight on noma

Abstract

Noma also known as Cancrum Oris is a destructive orofacial gangrenous disease which causes progressive and mutilating destruction of the infected tissues. It primarily affects under developed nations. The disease occurs mainly in malnourished young children with poor oral hygiene and debilitating concurrent illness. It is characterized by a rapid onset which usually starts in the mouth, spreads intra-orally destroying hard and soft tissue and progresses to perforate the facial skin leading to disfigurement. Treatment of acute noma includes transfusion of intravenous fluids, administration of broad spectrum antibiotics and debridement of necrotic areas. The main focus of the surgical treatment is to restore oral speech, oral competence and acceptable aesthetics by reconstructive surgery. Although, the mortality rate associated with noma has reduced significantly with the advent of modern generation antibiotics, the functional, cosmetic and psychological challenges associated with the demolition of soft and hard tissues still remains a problem. Noma is well documented in the literature, but because most patients do not report until the disease is at an advanced stage, its onset and progression is an enigma. The purpose of this article is to present a compendious review of the clinical course, aetiopathogenesis and treatment of noma to prevent and treat this devastating disease

Keywords: cancrum oris, gangrene, necrotizing stomatitis, noma

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Abbreviations: AIDS, acquired immunodeficiency syndrome; ANG, acute necrotizing gingivitis; EUSOL, edinburgh university solution of lime; ESR, erythrocyte sedimentation rate; HIV, human immunodeficiency virus; PKDL, post kala-azar dermal leishmaniasis; WHO, world health organization

Introduction

Noma or Cancrum oris is derived from the greek word nomein which means “to devour” or to graze.¹ This disease was described in 1848 by Tourdes as 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 parts and the bone and almost always quickly fatal”.² It is a necrotizing, destructive disease of orofacial tissues, and if not timely treated, it causes high morbidity and mortality. The introduction of modern antibiotics has changed the prognosis reducing the mortality rate from 90% to about 8%-10%.³

Noma is contemplated to portray the “face of poverty” because the 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, may lead to disease progression.⁴ The true incidence and prevalence of noma is unknown, because only 15% of patients with acute cases of the disease seek medical care and most patients with early stage noma are not reported to hospitals. As the clinical course of this disease is rapid most patients have advanced stage when they are reported to hospital. Noma is a disease of shame, so those affected are ignored rather than being taken for the medical care.⁵

Noma is truly an inexplicable disease and the understanding of it is confined. To incorporate the preventive measures and to develop

an early treatment regimen for noma, a better understanding of the aetiopathogenesis is required. The purpose of this article is to present a compendious review of the clinical course, aetiopathogenesis and treatment of noma to prevent and treat this distressing disease.

Discussion

History and epidemiology

Noma was first described by Hippocrates in 5th century B.C.⁶ It has been reported throughout history and from many parts of the world, primarily from poverty afflicted areas where the living conditions are dreadful. The disease was reported in Asia, Europe, South America, and Africa before the twentieth century but was almost eradicated with improvement in health care after the industrial revolution and elimination of poverty and malnourishment.^{2,7} Acute necrotizing gingivitis (ANG), an apparent precursor to noma, was commonly seen in the trenches on the western front in World War I and hence the name trench mouth originated for the disease.⁸ Noma was found in German and Japanese concentration camps in the course of World War II.⁷ In 1994, the World Health Organization (WHO) stated noma as a health priority and an action program was initiated by WHO, the United States National institute of health and the University of Maryland, Baltimore.⁹

The epidemiology of noma globally is unresolved.¹⁰ In the late 1990s, the incidence of acute childhood noma was set at 25,600 in countries bordering the Sahara.¹¹ and worldwide it's between 100,000 and 140,000 per year, unremarkably in sub-Saharan Africa and Asia.¹⁰ Peak incidence is among children aged 1-4 years.¹⁰ In 1997, worldwide extensiveness of those living with the sequelae of noma was placed at 770,000.⁵ Recent research indicates that noma is almost exclusively found in sub-Saharan Africa and in west Africa along the area running from Senegal across West Africa into Sudan and Ethiopia, coinciding with the so called “meningitis belt”.¹

Sporadic reports of solitary adult cases of noma seem to be a secondary disorder in individuals affected with complex diseases such as HIV/AIDS and other diseases.¹² Noma was also reported in patients with cyclic neutropenia, leukemia, Down's syndrome, Burkett's disease and herpetic stomatitis.¹³

Aetiopathogenesis

The exact etiology of Noma is not known but it is said to be multifactorial. It is described as a gangrenous disease in the literature. Gangrene by definition is an ischaemic necrotising process. But, the necrotic process in noma does not spread according to the distribution of the blood supply to the affected tissues, and thus noma is not primarily an ischaemic condition.¹⁴

Noma generally arises in conjunction with the presence of preexisting malnutrition, poor oral hygiene, and an inciting illness. Malnutrition induces alteration in cell-mediated immune response and breakdown of the epithelial tissues; alterations in the oral mucosa facilitate invasion by pathogens. Eating difficulties due to infection exacerbates the existing malnutrition. Stresses on malnourished children living in poor environmental conditions increase the level of circulating cortisol, which commences a cascade of reactions that impair the immune system and benefit the growth of bacteria.^{1,15}

It is difficult to identify the specific trigger agent in the complex microbiota of a noma lesion as it is an opportunistic infection. It has been contemplated that *Borrelia vincentii* and *Fusobacterium* (*Fusobacterium necrophorum* and *Fusobacterium nucleatum*) are prominent bacteria in such lesions.¹⁶ Symbiotic relationships between fusiform bacilli and non-hemolytic streptococci and staphylococci have been regarded as cardinal factors in the development of noma.¹ Recent data suggest that besides fusiform bacilli and spirochetes, other anaerobic bacteria are present in a relatively high proportion of noma lesions such as *Prevotella melaninogenica*, *Corynebacterium pyogenes*, *Bacteroides fragilis*, *Bacillus cereus*, *Corynebacterium pyogenes*, *Bacteroides fragilis*, *Bacillus cereus*, and *Prevotella intermedia*.⁶ *Fusobacterium necrophorum* is considered a prime component as this organism produces dermatotoxins, which could explain the rapid progression of the disease.¹⁷ *Fusobacterium necrophorum* is acquired by poverty stricken children through fecal contamination of water. *Prevotella intermedia* have the ability to break down lipid structures, which renders to tissue destruction. It also produces proteolytic enzymes which are highly capable of breaking down immunoglobulin G that ultimately hinders the elimination of microorganisms. 1 Few reports have suggested that these microorganisms are resistant to penicillin which accentuates the need for culture and sensitivity tests before administration of antibiotic.¹⁶

A recent study conducted by Hughie et al.¹⁸ contradicted the involvement of *Fusobacterium necrophorum* as an etiologic agent. Known periodontal pathogens like *aggregatibacter actinomycetemcomitans*, *capnocytophaga*, *porphyromonia* and *fusobacteria* were more prevalent in healthy samples compared to those with noma. Studies by Hughie et al.¹⁸ & Bolivar et al.¹⁹ identified *prevotella intermedia* and *peptostreptococcus* to be more clearly associated with noma. As it has a multifactorial etiology, additional studies are warranted to clarify its exact microbiology.

Clinical presentation

The clinical presentation of noma includes halitosis, excessive

salivation, severe dehydration and anemia.^{7,20} Systemic manifestations such as fever, tachycardia, lymphadenopathy, high respiratory rate, anorexia, general edema and ascites may represent an acute infectious process. Medical history reveals complain of parasitic or viral infection such as measles, malaria, recurrent fever and diarrhea. Blood examination reveals a low hemoglobin concentration and leukocytosis, elevated erythrocyte sedimentation rate (ESR) and hypoalbuminaemia.^{20,21}

The clinical presentation of noma is representative in most cases. Ninety percent of the cases occur before the age of 10 years with prevalence in the age of 1-4 years.⁷ A child is at an increased risk after the weaning period when the rich source of protein and protective antibodies are often replaced with a carbohydrate-rich diet and unsafe water, which may lead to malnourishment and diseases.²² Children with acute noma suffer from various stages of growth retardation and are adversely affected.²³

The World Health Organization has classified acute noma in four stages, ranging from halitosis with gingival bleeding to fulminant necrotizing stomatitis with tissue sloughing secondary to gangrene.²⁴ The very first recognized sign of noma is edema of cheek, or gingiva or both. A greyish black area appears on the external surface of the cheek contrary to the intraoral lesion which later on becomes a well-defined black necrotic zone. This necrotic zone acquires a cone shape and rapidly sloughs away.²⁰ Intra-oral manifestations include sequestration of the exposed bone and teeth, severe halitosis, pseudomembranes, extreme salivation, spontaneous gingival bleeding and loss of tips of interdental gingival papilla.^{6,20} At this stage the lesion is very painful with profuse salivation and strong malodor. The lesion is substantially larger on the inside than on the outside of the mouth involving both maxilla and mandible and it may also involve the temporomandibular joint, infraorbital margin, nose, antrum, and practically any part of the face. The gangrenous area is covered with bacterial masses and inflammatory cells. If no intervention is done, the process will persist and the child will die. It is estimated that between 70% and 90% of the children, if left untreated, will capitulate to complications from infection such as septicemia, pneumonia, or diarrhea. The child often is malnourished, apathetic and dehydrated and may suffer from underlying disorders such as malaria, HIV/AIDS, tuberculosis, or gastroenteritis.¹

Differential diagnosis

The differential diagnosis of noma includes leprosy; lupus erythematosus which is a slowly progressing disease; mucocutaneous leishmaniasis with disfiguring and destruction of nose, throat, and mouth; post Kala-azar dermal leishmaniasis (PKDL); oral cancer; syphilitic yaws (an infectious tropical disease caused by the bacterium *Treponema pertenue*); mucormycosis; agranulocytic ulcerations; physical trauma; chemical burns; acute herpetic gingivostomatitis; acute necrotizing ulcerative gingivitis (Vincent's disease, or trench mouth); clostridial or streptococcal gangrene; and lethal midline granuloma (also known as Stewart's granuloma or midfacial lymphoma).^{1,10,21} In the epoch of HIV/AIDS, opportunistic infections and diseases may make a differential diagnosis more challenging.¹

Noma neonatorum is a rare form of noma affecting newborns in the first months. It is a rare clinical syndrome leading to gangrene of orofacial tissues involving both term and preterm infants in the first week of life and is identical to noma in older children. The predisposing factors for noma neonatorum are preterm birth weight

and severe intrauterine growth retardation.^{6,20} Some authors believe that neonatal noma is a distinct entity; it was argued that noma neonatorum is a neonatal form of ecthyma gangrenosum and should not be classified as a form of noma.²⁵ Noma pudenda is another rare presentation of the disease which affects the anogenital (perineal) area and leads to necrosis of genitalia.^{1,25}

Risk factors

The most important risk factor for noma is poverty which itself breeds many other risk factors including chronic malnutrition, lack of education, poor environmental sanitation, poor oral hygiene, unsafe water, close living situations with domestic animals, exposure to animal and human fecal products, and exposure to viral and bacterial infections.²⁶

Among the major risk factors for noma, measles must be considered as one of the most menacing factors next to poverty. Severe measles is a terrible misery on children in the developing world and comparatively rare in the developed world because of compulsory child vaccination programs. Other diseases implicated in the pathogenesis of noma are malaria, tuberculosis, chicken pox, herpes infection, bronchopneumonia, and gastroenteritis.¹⁵

Although, HIV infection is not a strong risk factor, it may play considerable role in the pathogenesis of noma in South Africa. HIV-cytokine deregulation, HIV-induced depletion in the number of oral epithelial Langerhans cells and the HIV-associated dysfunction of immune regulatory cells may bring about oral tissue environmental conditions that permit bacterial colonisation, supporting the development of necrotizing diseases in the oral cavity.²⁷ It has been suggested that herpes viruses can also be a risk factor for noma. It is possible that upon activation, herpes viruses in cells of the periodontium and oral soft tissues, including keratinocytes, endothelial cells, fibroblasts and immuno-inflammatory cells may facilitate colonisation of anaerobic bacteria by deregulation of local immune mechanism and by inflation of local inflammatory responses. This results in increased risk of invasion by virulent anaerobic bacteria and tissue necrosis.²⁷

Complications

Mortality was a common complication of noma but with the use of modern antibiotics and better nutrition, mortality rate has been significantly reduced.²¹ Noma can result in trismus, sequestration of jaws, bony and fibrous ankylosis of temporomandibular joint, oronasal fistula, damage to permanent tooth bud, early loss of deciduous teeth and hypoplasia of maxilla and/or mandible. Most of the noma patients face difficulty in mastication because of hard and soft tissue loss. Severe cosmetic disfigurement can also take place from the resulting scarring and loss of tissue.¹³ Cases have been reported of a high psychiatric morbidity after noma. The resultant facial asymmetry increases with the growth of the child. The phrase "anarchie dentaire" is used to describe such features.² The infections from the oral cavity may even spread to other parts of the body.²¹ Other systemic complications such as dehydration, toxemia, and bronchopneumonia can occur and may lead to death of the child.²⁸

Treatment

The treatment can be divided into two major phases: intervention in the acute stage and surgical reconstruction. The treatment regimen must include broad spectrum antibiotics (ampicillin-cloxacillin and

metronidazole), analgesics, frequent use of anti-bacterial mouthwash chlorhexidine gluconate (0.12-0.2%) and gentle brushing to remove the superficial necrotic tissue and bacterial plaque. Fluid hydration (via nasogastric tube or intravenously), correction of electrolyte abnormalities, nutritional support, vitamin supplementation should be primarily given and patient should be put on high protein diet.^{20,21} The affected area should be debrided with dilute hydrogen peroxide or Edinburgh university solution of lime (EUSOL) or saline and any remaining tissue slough and sequester and any mobile tooth must be removed.^{13,21} Other diseases such as malaria, tuberculosis, parasites, and skin disorders should be addressed and treated. The patient must be screened for HIV infection and referred appropriately.^{1,6}

Surgical debridement and plastic reconstruction are usually necessary after severe tissue necrosis and sloughing occurs. After a period of disease quiescence of at least 6 to 18 months, surgical correction is initiated. The main focus of the surgical treatment is to restore oral speech, oral competence and decent aesthetics. Reconstructive surgery in children is delayed till the patient matures as it allows the defect to contract and reduce in size and ensures sufficient tissue for reconstruction.²⁹ Trismus resulted from extra-articular ankyloses (fibrosis) is corrected by complete excision of fibrosis followed by appropriate physiotherapy. Closure of tissue defects is usually done by various flaps namely local, pedicled or free flaps. Different other techniques employed include prefabricated scapular flaps, free radial forearm flap, anterolateral thigh flap, pedicled supraclavicular flap, waltzing flap and Gillies fan flap.¹³ Treatment done or not, a patient may perish due to debilitating underlying illness, systemic inflammatory response, malnutrition secondary to oral incontinence, aspiration pneumonia, or septicemia.²

Prevention

Noma is an uncommon disease; still clinicians should be aware of the early signs and symptoms of the disease, of its rapidly progressing course and of the hazards of delay in seeking professional advice and treatment; as the consequences of disease are grave and fatal. Public health department should incorporate the measures for routine screening of malnourished children in underprivileged societies who are at risk of this disease and making available immediate and effective treatment for them. Various measures to prevent noma include administration of nutritious food, exclusive breast feeding upto six months of life, inculcation of proper oral hygiene practices, immunization against endemic diseases like measles, segregation of animals from human living areas and creating cognizance about noma among the poor population of the world.^{14,20}

Conclusion

Noma or Cancrum oris is a devastating disease primarily affecting the poor and young children. It generally occurs among the underprivileged social environments where even primary medical care is not available. It can be prevented to a large extent by spreading proper education of the disease about its causes and manifestations to the poor population; by providing good nutrition and water facilities, vaccinations and by maintaining proper hygiene. The government and health organizations must take suitable steps to improve the social living conditions of individuals living in noma susceptible areas. Efforts should be made by medical professionals and health care providers to provide early intervention and medical care to the patients so that mortality and morbidity can be reduced. Recent updates on the causes of the disease and nature of its progression, summarized in this

review, may guide the clinician in better managing this debilitating disease. Efforts must be taken globally to eradicate this disease.

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Conflict of interest

The author declares no conflicts of interest.

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