

CANCRUM ORIS - A DESCRIPTIVE CASE SERIES ON 13 PATIENTS

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ABSTRACT

Objectives: To evaluate the age, gender, site distribution and associated systemic debilitating conditions (if any) of the patients with Cancrum Oris.

Methodology: A descriptive retrospective study was undertaken in the Oral and Maxillofacial Surgery unit of Khyber College of Dentistry from January 2005 to January 2010. Information regarding the variables of the study i.e., age, gender, site distribution and associated systemic debilitating conditions, was collected from the ward charts during the study period.

Results: A total of 13 patients reported to the unit during the study period, 8 males and 5 females with male to female ratio of 1.6:1. The age range of the patient was from 9 to 70 years with the mean age of 37.69 SD + 22.65. The first and the third decade of life was the most common age group affected with Cancrum Oris i.e., 23.07% each, with another peak in the sixth decade of life i.e., 23.07%. Bilateral anterior maxilla was affected in 30.76% cases. Uncontrolled diabetes was the common associated systemic debilitating condition i.e., 72%. All of the patients were from low socio-economic class.

Conclusion: Cancrum oris is a disease of low-socioeconomic group. In our series of patients the most common associated debilitating systemic condition was diabetes mellitus with a bimodal age distribution.

Key Words: Cancrum Oris, Anterior maxilla, Uncontrolled diabetes, Debilitating conditions.

INTRODUCTION

Cancrum Oris or Noma has been referred to as “maladie dévoreuse de beauté et de vie” (“An illness devouring beauty and life”) by Edmond Kaiser, founder of the humanitarian organization Sentinelles. The word originates from the Greek verb *numein* meaning “to devour”. This gangrenous disease affects both soft and hard tissues of the mouth and face. It occurs predominantly in children of less developed countries

having poor hygienic conditions and debilitating diseases. This is a disfiguring disease, and thus it has also been named “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 contribute to disease progression.^{1,2}

The etiology of cancrum oris is not known, but it is assumed to be infectious in origin. Some of the risk factors include low socioeconomics; malaria, malnutrition, measles, and poor oral hygiene.³ There are several forms of the Cancrum Oris (Noma). The classic form of the disease mostly affects children be-

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tween 3 and 6 years of age in under developed countries. Noma of the debilitated adult is another form, which is less locally invasive, and occurs in immunocompromised adults with major debilitating diseases in both developed and developing countries. Noma neonatorum, is yet another form of the disease, has been called Noma due to similarity of the facial lesions but it is a different disease which affect premature babies. In this form in addition to destruction of the oral and peri-oral tissues there is necrosis of the perineal region, which is highly fatal because of irreversible septicaemia. It is important to remember that Noma neonatorum is a discrete pathological entity that should not be confused with the classic form of Noma.^{4,5}

Cancrum oris was first described by Tourdes as a gangrenous affectation of the mouth. It was known as early as the time of Hippocrates.⁶ 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.^{7,8} The World Health Organization has reported alarming increases in the incidence of Noma over the past 10 years.^{2,9}

In Noma, the specific trigger agent in the complex microbiota is difficult to identify but it has been speculated that *Borrelia vincentii* and *Fusobacterium* are important bacteria in such lesions.^{5,10} Symbiotic relationships between *Fusiform bacilli* and non-hemolytic *Streptococci* and *Staphylococci* have been considered significant factors in the causation of Noma. Lack of specimens from early stages of the lesion, confusing microbial taxonomy, lack of animal models for studies and difficulty in culturing microorganisms were a few problems in the previous studies.¹⁰ Recent reports show that besides *Fusiform bacilli* and *Spirochetes*, other anaerobic bacteria are present in a relatively high proportion of Noma lesions.⁵ A key component is *Fusobacterium necrophorum*, this organism produces a toxin called dermatotoxins, which could explain the rapid progression of the disease.¹¹ *Fusobacterium necrophorum* is acquired by impoverished children through fecal contamination of water, which occurs when residential facilities are shared with animals and sanitation is very poor. A major contribution in tissue destruction is caused by *Prevotella intermedia* which has the ability to break down lipid structures. This microbe also produces proteolytic enzymes capable of breaking down Immunoglobulin G, which impedes the elimination of microorganisms. Some studies suggested that these

microorganisms are resistant to penicillin,⁵ which emphasizes the need for culture and sensitivity tests before administration of antibiotics. Additional studies are warranted to elucidate its exact microbiology because some authors suggest a multifactorial etiology for this condition.¹²

METHODOLOGY

This descriptive study was conducted at the Department of Oral and Maxillofacial Surgery Khyber College of Dentistry, Peshawar from January 2005 to January 2010. The Department of Oral and Maxillofacial Surgery of Khyber College of Dentistry is a tertiary care center of Khyber Pakhtunkhwa Province of Pakistan. This unit receives patients from entire Province as well as from some parts of Afghanistan and Federally Administered Tribal Areas (FATA). All the patients reporting to the Department of Oral and Maxillofacial Surgery with Cancrum Oris were included in the study. Information regarding the variables of the study i.e., age, gender, site distribution and associated systemic debilitating conditions was collected from the ward charts during the study period. Plane X-rays like Orthopantomogram (OPG), Postero-anterior view face, Paranasal sinus view (PNS), right and left Lateral Oblique views of the mandible and advance imaging like Computed tomographs (CT Scans) were used for the diagnosis of patients with Cancrum Oris and to know the exact extent of the disease.

RESULTS

The present study showed that Cancrum Oris is more common in males (62%) as compared to females (38%) with a male to female ratio of 1.6:1. Our series of patients showed a bimodal age distribution with the first peak in first and third decade of life and the second peak in 6th decade of life, i.e., 23.07% each. The mean age of these patients was 37.69+22.65 years (Table 1). Bilateral anterior maxilla was affected in 30.76% cases. The involvement of other sites of oral cavity is given in Table 2. The most commonly associated systemic debilitating condition was uncontrolled Diabetes i.e., 72%, while Hepatitis C viral infection and Valvular heart disease were the other two associated systemic condition constituting 14% each (Figure 1).

DISCUSSION

Cancrum Oris or Noma tracks a typical course of disease progression. First the oral mucosa of the

Table 1: Age Distribution

Age of the patients in years	Number of patients	Percentage
1 to 10	3	23.07
21 to 30	3	23.07
31 to 40	1	7.69
41 to 50	1	7.69
51 to 61	3	23.07
61 to 70	2	15.38
Total	13	100

Table 2: Site Distribution

Age of the patients in years	Number of patients	Percentage
Premaxilla and upper lip	1	7.69
Bilateral anterior maxilla	4	30.76
Posterior hard palate	2	15.38
Lower lip and alveolus	2	15.38
The entire maxilla	2	15.38
Maxillar Tuberosity	2	15.38
Total	13	100

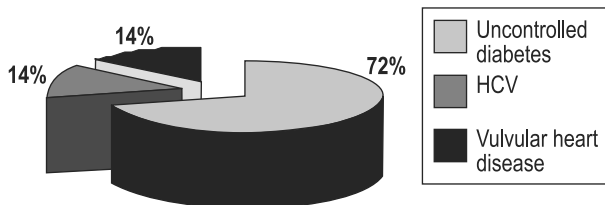


Fig. 1: Associated systemic diseases in patients with cancrum oris

gums, lining of cheeks become inflamed and develop an ulcer. The infection spreads from the oral mucosa to the skin thus causing necrosis of the tissues of lips and cheeks.¹³ There is sore mouth with focal edema having fetid odour and taste. Foul smell and purulent oral discharge is associated with copious salivation, anorexia and palpable cervical lymphadenopathy. First there appears a small, tender purplish red spot on the gums, which quickly becomes indurated, ulcerated and then becomes necrotic with associated edema. It forms a bluish black necrotic cone-shaped mass with intra-oral base. There is rapid progression to gangrene during the next 4 to 72 hours. It may produce extensive facial disfigurement with loss of intraoral structures

and functions. Rapid, painless tissue break down continues and can progress to destroy the soft tissues and even the bone. Occasionally the condition involves genitals and is called Noma Pudenda.^{13,14}

Although Noma is witnessed in adults,¹⁵ the disease typically affects mostly children between 3 and 12 years of age,^{16,17} but according to the present study Cancrum Oris has shown a bimodal age distribution with the first peak in the 1st and 3rd decade of life and the second in 6th decade of life. A background of poor oral and general hygiene characterizes children at risk for this disease, and most cases of Noma present with a distinct history of recent prior debilitating infections, with measles being the most frequent.^{18,19} Other antecedent infections reported in 69 Noma cases by Enwonwu¹⁸ included malaria (30%), chicken pox (9%), tuberculosis and others (17%). It must be emphasized that in impoverished African children, single infections are very rarely encountered. Measles infection is known to down-regulate Inter-leukin-12 (IL- 12), which is needed to generate cell-mediated immunity,²⁰ and studies have shown that malnourished children with compromised vitamin A status are prone to develop deep erosive ulcers of the mouth and eyes following measles attack^{21,22}

According to Obiechin and Aribota²³ (52.6%) of the patients were males and (47.4%) females. However in our study, this disfiguring condition is more common in males as compared to female with the male to female ratio of 1.6:1.

The involvement can be uni- or bilateral and may affect any part of the face including upper/lower jaw.^{13,14} Bilateral anterior maxilla was the most common site for patients with cancrum Oris i.e., 30.67% according to the present study.

Noma is preceded most of the time by predisposing illnesses such as measles, tuberculosis, leukemia and AIDS.^{24,25} An increasing incidence of Noma and noma-like lesions in people with AIDS have also been reported.²⁶ Some of the Noma case reports shows bronchiectasis, as well as cystic fibrosis, which affects the pancreas and intestinal tract, causing intestinal obstruction and malabsorption;²⁷ 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.² Eating difficulties due to infec-

tion 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.^{2,9}

According to the present study the most common debilitating systemic illness associated with Noma was uncontrolled Diabetes Mellitus constituting 72% of the patients, though the sample size of the study was very small. Only thirteen patients reported during the study period which indicates the rare occurrence of this entity in the region. These patients often suffer from fever, tachycardia, tachypnea and anorexia. The disease is rapidly fatal if left untreated.²⁸

The management of noma requires a multidisciplinary team approach. In the early stages, the child will need oral irrigation with hydrogen peroxide, saline and 0.2% chlorhexidine, which helps to slough the necrotic tissue. Hydration, correction of electrolytes and vitamin deficiencies with provision of sufficient nutritional support is essential, even through nasogastric tube, if necessary. There is no clear agreement but most authors recommend Penicillin plus Metronidazole to cover principal organisms. Medication needs to be continued for at least 14 days. The favored antibiotic is Penicillin G 2.4 million U IV QID + Metronidazole 500mg IV TDS. The other option is Ampicillin/Sulbactam 3.0 g IV 6 hourly. There may be overgrowth of *Candida* due to the use of antibiotics, thus requires antifungal coverage with Nystatin rinses 5ml QID or Fluconazole 200mg orally once daily. Improvement of nutrition, cleanliness and sanitation and early vaccination is required to prevent this disfiguring condition.²⁹ Plastic and reconstructive surgery is required in most of the cases at later stages, because functional disturbances and disfigurement are the hallmark of the disease.³⁰

CONCLUSIONS

1. Our series of patients showed a bimodal age distribution.
2. The condition is common in males as compared to females.
3. Bilateral anterior maxilla was the most common site affected by the disease.
4. Uncontrolled Diabetes was the most common associated systemic condition in patients with Noma.

5. Facial disfigurement is the hallmark of the condition that is why early diagnosis and Multi-disciplinary approach is required to combat this disfiguring condition.

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