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Short Communication

Noma (Cancrumoris) in the Western World

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Abstract

Noma is a devastating orofacial gangrenous polymicrobial infection, which is predominantly seen in malnourished children in developing (especially sub-Saharan) countries. The pathogenesis is multifactorial and thought to result from a complex interaction between opportunistic, bacterial pathogens, poor oral hygiene, and malnutrition in patients debilitated by severe illness or immunosuppression. The disease is associated with high morbidity and mortality. Only few cases of noma have been described in the Western world and to our knowledge noma have not previously been reported in Scandinavia. We describe a 37-year-old Caucasian male admitted due to acute necrotizing stomatitis advancing to fulminant noma within few days. The case report illustrates that noma should be considered as a differential diagnosis in patients presenting with severe orofacial infections, even in the Western world.

ABBREVIATIONS

ENT: Ear-Nose-Throat

INTRODUCTION

Noma, also known as cancrumoris, is a devastating, progressive, orofacial gangrenous disease.

It is predominantly seen in malnourished children in the developing countries. Affected children typically suffer from a combination of debilitating illness and poor oral hygiene, which predisposes to oral infections [1,2]. Noma starts as an acute gingivitis. Left untreated, the gingivitis may advance to necrotizing periodontitis. The soft and hard tissues of the oral cavity (necrotizing stomatitis) and the facial structures are affected in later phases [3,4]. Well-demarcated full thickness skin destructions and exposed bone with exfoliation of teeth are other characteristics of noma [5]. Untreated noma is generally lethal.

Within the last century, only few cases of noma arising in the Western world have been reported in the literature [2-3] and to our knowledge, noma has not previously been described in Scandinavia. We present a 37-year-old Caucasian male admitted due to severe necrotizing gingivitis progressing to fulminant noma within a few days.

CASE PRESENTATION

A 37-year-old Caucasian man with a large daily alcohol intake was admitted to the local hospital due to somnolence and abdominalia on June 14th 2014. Blood tests revealed electrolyte derangement and signs of acute pancreatitis (Table 1).

Due to the development of trismus and difficulty swallowing the patient was seen by an Ear-Nose-Throat (ENT) specialist on June 17th. Physical examination revealed necrosis involvingthe inferior gingival mucosa, the alveololabial and alveolobuccal sulci extending into the retromolar regions, the floor of the mouth, the right side of the tongue, and the skin of the lower lip. The patient refused further treatment and left the hospital.

Two days later, the patient was re-admitted, because he was unable to eat and drink. The extent of the necrosis had increased. At this time he had a full-thickness necrosis and perforation of the lower lip (Figure 1). Trismus, intense halitosis, and excessive salivation were noted. Antibiotic treatment with intravenous cefuroxime and metronidazole was initiated. The following day the patient was transferred to the ENT-department, Aarhus University Hospital, where the patient underwent surgical debridement immediately. The right half of the tongue, the anterior part of the floor of the mouth, the mucosa of the hard

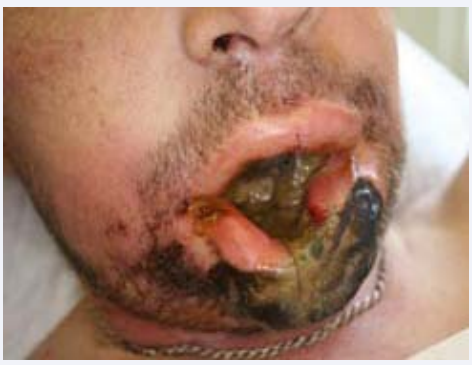

Figure 1 Photograph taken upon admission.

Table 1: Selected biochemical test results from blood samples obtained during the period June 14th to June 23rd.

	June 14th	June 17th	June 20th	June 23rd	Reference levels
C-reactive protein (mg/l)	108	218	241	74	< 8
Leucocyte count (x 10^9/l)	13.4	26.2	14.7	14.4	3.5 - 10
Neutrophil count (x 10^9/l)	12.0	24.1	11.9	10.4	2 - 7
Hemoglobin (mmol/l)	7.9	7.0	4.9	5.5	8.3 - 10.5
Creatinine (µmol/l)	174	184	264	76	60 - 105
Sodium (mmol/l)	117	131	147	152	137 - 145
Potassium (mmol/l)	2.6	3.1	3.2	4.6	3.5 - 4.6
Amylases (U/l)	818			451	10-65
Albumin (g/L)	31	28	16	14	36 - 48

palate, superior and inferior gingiva, and inferior lip were necrotic and removed. Furthermore, several teeth were loose and removed (Figure 2). No signs of infection on the neck were found. The antibiotic treatment regimen was changed to intravenous meropenem, clindamycin and ciprofloxacin.

Surgical revision and debridement was performed daily the following days. On June 23rd large parts of the mandible and maxilla were necrotic and removed. Nude parts of the mandible and maxilla were treated with cod-liver oil ointment and an overlying non-medicated paraffin dressing [6]. Subsequently, the infection subsided and the mucosal lining of the oral cavity was re-established. However, severe sequelae remained: the lacking lower lip impaired retention of saliva, therapy-resistant extreme trismus in combination with loss of more than half of the tongue and parts of the palate made the speech indistinct and impaired oral intake of food and liquid.

Seven months after the onset of symptoms, the patient underwent reconstructive surgery including recreation of the lower lip using bilateral karapandzic flaps, tongue plastic with split skin transplant, and bilateral rigottomy of the mandibular rami with lipo-transplantation.

One year after presentation, oral continence and speech are much improved, but the refractory trismus prevents dental rehabilitation. These problems cause continued impaired oral intake. Bacterial cultures from oral necrotic tissues revealed growth of Viridans streptococci (++++), Bifidobacterium species (+++), Lactobacillus species (+++), Candida albicans (++), and coagulase-negative Staphylococci (++). No growth was detected in blood cultures.

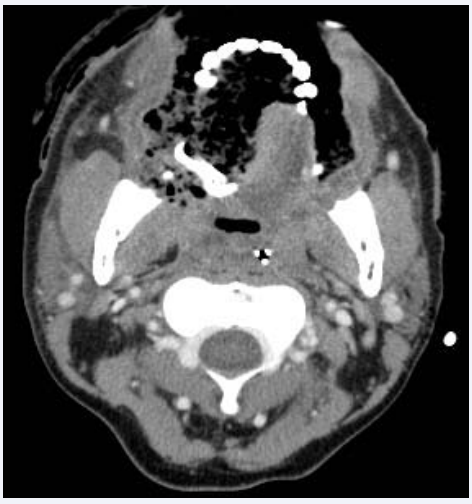

Figure 2 Computed tomography showing the extent of infection.

Histological examination of the removed oral tissue showed acute infection and necrosis without signs of malignancy (Table 1). Selected biochemical test results from blood samples obtained during the period June 14th to June 23rd.

DISCUSSION

Noma was first described by, Hippocrates in the 5th century BC. In 1848, Tourdes gave a definition of noma, which is still valid today: "a gangrenous disease affecting the mouth and face of children in bad hygiene conditions and suffering from debilitating disease, especially eruptive fever, beginning with an ulcer on the oral mucosa rapidly spreading outside and destroying the soft and hard tissues of the face – and almost always fatal" [2].

Noma was common in Europe and USA in previous centuries [5,7]. Along with improvement in the standard of living and decrease in extreme poverty and starvation, noma disappeared from the Western world in the 20th century [5,7]. The disease reappeared in concentration camps during World War II, and more recently it has been described in rare case reports froma few developed countries. In these cases noma has been associated with debilitating disease, intense immunosuppression, and malnutrition in combination with poor oral hygiene [3,5]. On the contrary, nomais still prevalent among malnourished young children (1-4 years old) in developing countries, especially in the sub-Saharan region [1,3,7].

Noma is typically initiated by a gingival ulceration, which progresses to necrotizing gingivitis. Left untreated, the infection may advance to necrotizing periodontitis and stomatitis involving most structures of the oral cavity. The infection spreads rapidly through both soft and hard orofacial tissues and a characteristic well-defined blackened necrotic centre is typically seen. Sequestration of exposed bone and teeth occurs spontaneously [2-3,5]. Perforation of the cheek is seen in the majority of cases along with extensive intraoral destructions [2,5].

The clinical symptoms and findings include severe pain, fever,

Table 2: Described cases of noma in the Western world.

Country and year of publication	Patient age (years)	Debilitatingdisease	Mal-nutrition	Anatomic site	Microorganisms	Reconstructivesurgery
USA, 1974 [11]	27	Leukaemiapyelonephritis	NS	Chin Oral and nasal cavity	Alpha-hemolyticstreptococci *Neiseeriacatarrhalis Proteus mirabilis*	Not performed (patient died)
USA, 1976 [12]	63	Leukaemia	NS	Lip oral cavity	Gram-positivediplococcic Gram negative rods *Bacteroidesmelaninogenicus*	Not performed (patient died)
USA, 1995 [8]	46	HIV AlcoholismIntravenous drug abuse	Yes	Chin Oral cavity	Gram-negative rods Gram-negativecocci in pairs and chains *Staphylococcusaureus*	Regional tissue flaps
Wales, 2001 [7]	70	Diabetes mellitus	No	Lips Oral cavity	Beta-hemolyticstreptococci *Haemophilusinfluenzae Escherichiacoli*	Not described in details
France, 2002 [9]	31	AIDS Non-Hodgkinslymphoma	NS	Lips Chin Oral cavity	Anaerobic rods Coagulase negative staphylococci *Enterobacteraerogenes Pseudomonasaeruginosa*	Not performed (patient died)
USA, 2003 [14]	65	Oral myiasis Acuterespiratory-failure	Yes	Lip Chin Oral and nasal cavity	Not described in details	Not performed (patient died)
United Kingdom, 2006 [13]	68	Hypothyroi-dism Hepatitis B	Yes	Oral cavity	Not described in details	Not described in details
Italy, 2009 [4]	89	NS	Yes	Chin Oral cavity	Mixed skin flora withoutsignificantgrowth	Not performed (patient died)
USA, 2011 [10]	40	HIV/AIDS, Hepatitis B	Yes	Chin Oral cavity Neck	*Prevotella species Peptostreptococcus species*	Not described in details

Abbreviations: NS: Not specified. HIV: Human Immuno-deficiency Virus. AIDS: Acquired Immune Deficiency Syndrome.

gingival ulceration, trismus, mucosal swelling, purulent discharge, excessive salivation, and extreme halitosis. Lymphadenopathy, leucocytosis, hypoalbuminaemia, and anaemia are typically described [2-3,5].

The course of the disease is rapid, and death can occur few days after the onset of mucosal oedema [2]. Without treatment, the majority of patients will die within few weeks after the debut of symptoms [2,7].

Those who survive noma often suffer from severe sequelae including facial disfigurement and functional impairment including displacement of the teeth, trismus, oral incontinence, nasal regurgitation, and impaired speech [1,5,7].

The pathogenesis of noma is thought to be multifactorial [5]. Essential factors for the development of noma include a polymicrobial oral infection in a malnourished and immunosuppressed individual, who is often in a state of debilitation due to systemic infection [5]. Measles and malaria are the most frequent preceding debilitating diseases in patients from the developing countries [3,5]. In the Western world, rare case reports of noma (in adults) have been associated with different coexisting, debilitating diseases such as hepatitis B, leukaemia, and Acquired Immune Deficiency Syndrome (AIDS) (Table 2) [4,8-15]. A combination of malnutrition, debilitating disease and oral infection is described in most cases. Cultures from oral cavity specimens have shown a wide variety of microorganisms and there is no consensus regarding the causative microorganisms in noma [2-3]. *Prevotellaintermedia* and *Fusobacterium necrophorum* have been recovered from noma lesions and some researchers believe they are key-pathogens [2-3]. Herpes viruses may play an initiating role, as this virus infection has been shown to impair local immunity and thus facilitate infection with pathogenic bacteria derived from the oral flora [5]. However, the diagnosis of noma is based on the medical history and the clinical presentation rather than the microbiological findings. In previous case reports, patients were treated with broad-spectrum antibiotics, surgical revision, and daily debridement. The majority of patients died in the acute phase and reconstructive procedures were not performed.

The differential diagnoses of noma include necrotizing fasciitis, a granulocytic angina, malignant lesions (including extra nodal natural killer/T-cell lymphoma), leishmaniasis, yaws, and syphilis [5,9,11]. However, most authors agree that the diagnosis is undisputable when the described classic symptoms and objective findings are present [9].

In order to achieve preferable results, immediate and aggressive treatment is crucial. Broad-spectrum antibiotic treatment covering the oral florain combination with

necrotectomy and wound-care is recommended [2]. Furthermore, nutritional rehabilitation, correction of dehydration and electrolyte imbalance, and treatment of predisposing diseases is also important [3]. Even in cases treated promptly and correctly, noma typically leaves behind severe facial disfigurement and functional impairment. Hence, most patients will need advanced reconstructive plastic surgery in later phases [2-3].

We believe, that the described patient represents a case of noma, because: (1) He presented with acute, necrotizing stomatitis, which progressed to a necrotic, blackish full-thickness lower lip destruction within a few days, (2) The infection spread rapidly through anatomic barriers including skin and bone, (3) The patient was malnourished due to alcoholism and debilitated due to acute pancreatitis, (4)Highly elevated inflammation markers and fever indicated infection (Table 1), and (5) Oral cultures showed polymicrobial growth.

The current case illustrates that noma can develop in the western world when certain conditions coexist and clinicians should be aware of the characteristic clinical presentation in order to initiate life saving immediate and aggressive treatment.

REFERENCES

1. Marck KW. Cancrum oris and noma: some etymological and historical remarks. Br J Plast Surg. 2003; 56: 524-527.
2. Baratti-Mayer D, Pittet B, Montandon D, Bolivar I, Bornand JE, Hugonnet S, Jaquinet A. Noma: an "infectious" disease of unknown aetiology. Lancet Infect Dis. 2003; 3: 419-431.
3. Enwonwu CO. Noma--the ulcer of extreme poverty. N Engl J Med. 2006; 354: 221-224.
4. Chiandussi S, Luzzati R, Tirelli G, Di Lenarda R, Biasotto M. Cancrum oris in developed countries. Aging Clin Exp Res. 2009; 21: 475-477.
5. Enwonwu CO, Falkler WA Jr, Phillips RS. Noma (cancrum oris). Lancet. 2006; 368: 147-156.
6. Terkelsen LH, Eskild-Jensen A, Kjeldsen H, Barker JH, Hjortdal VE. Topical application of cod liver oil ointment accelerates wound healing: an experimental study in wounds in the ears of hairless mice. Scand J Plast Reconstr Surg Hand Surg. 2000; 34: 15-20.
7. Marck KW. A history of noma, the "Face of Poverty". Plast Reconstr Surg. 2003; 111: 1702-1707.
8. Evans LM, Lane H, Jones MK. Cancrum oris in a Caucasian male with Type 2 diabetes mellitus. Diabet Med. 2001; 18: 246-248.
9. Barrios TJ, Aria AA, Brahney C. Cancrum oris in an HIV-positive patient. J Oral Maxillofac Surg. 1995; 53: 851-855.
10. Mayorca A, Hazime N, Dekeister C, Paoli JR. Necrotizing stomatitis after radiotherapy in a patient with AIDS: case report. J Oral Maxillofac Surg. 2002; 60: 100-101.
11. Silva KN, Twaddell WS, Powers DB. A 40-year-old man with a perforated cheek. Am J Med Sci. 2011; 341: 399-403.
12. Weinstein RA, Choukas NC, Wood WS. Cancrum oris-like lesion associated with acute myelogenous leukemia. Oral Surg Oral Med Oral Pathol. 1974; 38: 10-14.
13. Limongelli WA, Clark MS, Williams AC. Nomalike lesion in a patient with chronic lymphocytic leukemia. Review of the literature and report of a case. Oral Surg Oral Med Oral Pathol. 1976; 41: 40-51.
14. Buchanan JA, Cedro M, Mirdin A, Joseph T, Porter SR, Hodgson TA. Necrotizing stomatitis in the developed world. Clin Exp Dermatol. 2006; 31: 372-374.
15. Aguiar AM, Enwonwu CO, Pires FR. Noma (cancrum oris) associated with oral myiasis in an adult. Oral Dis. 2003; 9: 158-159.

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