Noma management in a 47-years-old female patient using estlander flap: A case report

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ABSTRACT

Background: Noma or cancrum oris is an uncommon disease that results in loss of tissue in the oronasal region secondary to gangrenous slough. As most patients with noma do not report until the disease is at an advanced stage, its onset and progression remain a mystery. Survivors of this disease suffer severe facial deformity due to loss of facial tissues and scarring. Reconstruction of such type of defects was aimed to maintain oral competence, sufficient oral access, and preservation of sensation.

Case: A woman, 47 years old, experience with a chief complaint of the deformed right upper lip for the past 45 years. The patient is known to have a history of deformity on the right upper lip region when the patient was two years old. The patient stated that it originally started

with swelling on the right cheek accompanied by redness and painful sensation.

Result: An extensive scar along with half of the upper lip missing was found in the physical examination, causing exposure of anterior maxillary teeth. A full-thickness defect of half of the upper lip with severe scar contracture was also found in the further examination, thereby constricting the oris. The patient was planned for surgical management under general anesthesia. Lip reconstruction using an Estlander flap was done.

Conclusion: Reconstruction must be planned carefully to restore the natural contours of the lip. The Estlander flap is one of the treatment choices used for labial defects that include the commissure.

Keyword: noma, facial deformity, Estlander flap

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INTRODUCTION

Noma or in other name cancrum oris, fusospirochetal gangrene, necrotizing ulcerative stomatitis, or stomatitis gangrenosa is an old companion of humankind. It refers to an uncommon disease that results in loss of tissue in the oronasal region secondary to gangrenous slough and has been known since the time of Hippocrates, and Galen.¹ Noma is a rapidly progressive polymicrobial opportunistic infection resulting in orofacial gangrene and eventually death if untreated. The etiopathogenesis of the disease has been attributed to extreme malnutrition, dehydration, and inadequate oral hygiene.^{1,2}

In previous centuries the disease has been described commonly in European countries and the USA.³ It disappeared from these countries due to increased welfare, enabling even the poorest to feed their children adequately. The disease has been forgotten and disappeared almost entirely from modern medical textbooks. However, noma is still present in countries where extreme poverty and malnutrition are prevalent.

CASE REPORT

A 47-years-old female patient came to the surgical clinic of Dharma Yadnya General Hospital with a

chief complaint of the deformed right upper lip for the past 45 years. The patient is known to have a history of deformity on the right upper lip region when the patient was two years old. Based on the interview, the patient stated that it originally started with swelling on the right cheek accompanied by redness and painful sensation. As time goes by, the patient noticed that an open wound was showing on the right upper lip region, which eventually turned into an ulcer accompanied by pus and finally turned into gangrene. The patient didn't make any effort to seek treatment even until that point due to economic reasons. She has decided to seek treatment now because the patient has been getting help in the form of funds from a local foundation. The patient is known to come from a rural region in Bali where the economic status is below average, and patient's family was faced with difficulties regarding food availability, forcing the patient to consume foods less than the patient's daily nutrition requirements. An extensive scar, along with half of the upper lip missing, was found in the physical examination, causing exposure of anterior maxillary teeth (Figure 1). A full-thickness defect of half of the upper lip with severe scar contracture was also found in the further examination, thereby constricting the oris. The patient was planned for surgical management under general anesthesia. Lip

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Figure 1 Frontal views of the patient showing Noma

Figure 2 Estlander Flap Design

reconstruction using an Estlander flap was done after the patient's blood test and thorax photo result within the normal range (Figure 2). Incision markings were made, and local anesthesia was injected for local hemostasis. The flap was raised and mobilized, dissected carefully up to vermillion on the pedicle side of the flap, and care was taken not to expose labial vessel, the entire flap was rotated 90° into the defect (Figure 3).

DISCUSSION

As the old companion of humankind, noma is still commonly seen in a population with extreme poverty, severe malnutrition, unsafe drinking water, poor sanitation, poor oral health practices, high infant mortality, limited access to high-quality health care, and intrauterine growth retardation.³

Figure 3 Intraoperative photograph showing the rotation of the Flap after final suturing

Recently, an increased incidence of noma has been reported in patients with Human Immune deficiency Virus (HIV) infection.⁴ malnutrition is considered to be an important risk factor for noma. In Africa, most of the cases were reported during the dry season when food is scarce, and when the incidence of measles is highest.⁵ host resistance, debilitating diseases like malaria and measles were considered to be significant risk factors or precursors to noma. Measles could be an essential risk factor because of the associated immunosuppression.⁴ The mortality rate of noma is not well known. Historical data indicate that 80–90% of noma patients die due to sepsis. The mortality rate can decrease significantly to around 10–20% if the patient receives medical attention at an early stage of the disease and treated with oral antibiotics. However, most noma patients do not have access to healthcare.⁵

The exact etiology of noma is not known, but it is believed to be multifactorial in nature. Researchers initially thought that bone exposure caused by acute necrotizing ulcerative gingivitis (ANUG) could act as a passage point for noma. But the present consensus shows that ANUG is a precursor for noma.⁴ It is suggested that some factors such as malnutrition, weakened immune functions, and prior viral infection, all worsened by inadequate oral hygiene, could lead to a reduction in host resistance and favor the development of oral ulcers. These lesions can serve as entry sites for microorganisms responsible for the disease process.⁶ When poverty prevails, there appears to be a synergistic relation between malnutrition, weakened immune functions, and increased susceptibility to infections. Immunoglobulin A, an important component

of the mucosal immune system, is significantly reduced. In contrast, the plasma concentration of pro-inflammatory cytokines and C-reactive protein is increased in malnourished subjects compared to healthy counterparts. Infections such as AIDS, measles, and malaria could also lead to a shift from pro-inflammatory cytokines to anti-inflammatory cytokines.⁵

Rapid destruction of hard and soft tissues seen in noma could be attributed to immunopathological response to microbial factors rather than microbiological factors alone. Enwonwu et al. found higher plasma levels of anti-inflammatory and pro-inflammatory cytokines in children with necrotizing ulcerative gingivitis when compared to controls.⁵ Another research has shown that oral epithelial cells and other resident cells secrete several pro-inflammatory cytokines and chemokines in response to bacterial products, which in turn can stimulate the expression of matrix metalloproteinase.⁶ This matrix metalloproteinase can cause the destruction of both hard and soft tissues.

Noma is an opportunistic infection; the role of specific microorganisms in the pathogenesis has not been explained. A number of potential pathogens were found in abundance in the sites of noma which include *Prevotella melaninogenica*, *Corynebacterium pyogenes*, *Fusobacterium nucleatum*, *Bacteroides fragilis*, *Bacillus cereus*, *Prevotella intermedia*, and *Fusobacterium necrophorum*.⁴ Microbial analysis in the early 20th century revealed the presence of spirilliform and fusiform microorganisms in biopsy samples taken from the transitional zone between gangrenous and healthy tissues.⁶ Later studies reported that *Fusobacterium necrophorum*, a predominant animal pathogen to be the most common microorganism isolated from the disease sites in Nigerian children. It was suggested that *Fusobacterium necrophorum* could be a trigger organism for noma. This microorganism produces various toxins and has been associated with necrotizing infections in animals, and it may contaminate livestock and potentially infect children.^{4,5}

The clinical course of noma starts with facial edema, extreme halitosis, and hypersalivation, where intraoral inspection may reveal a painful red spot or a necrotizing gingivitis/stomatitis; Within a few days, a bluish discoloration appears in the edematous region, after which the skin breaks down over the extensive underlying necrosis; After separation of the gangrenous tissue, a large hole result often showing devitalized bone that will sequester at a later stage; If the patient survives, natural healing frequently leads to a mutilated face with severe functional impairments like trismus or ankylosis

of the mandible, drinking, eating and speech problems, oral incontinence, and sequelae often result in social isolation.^{7,8,9,10}

Reconstruction must be planned carefully and executed meticulously to restore the natural contours of the affected area. The functional goals of lip reconstruction include maintenance of oral competence, sufficient oral access, and preservation of sensation. Aesthetically, facial units should be reconstructed with adequate tissue match in terms of color and texture, aiming at symmetry as well as the preservation of the apparent commissure and philtral structures.¹¹ The highest degree of aesthetic camouflage in lip reconstruction is obtained by primary closure and with adjacent or local flaps. Estlander flap is one kind of local flap that can be used for labial defects, including the commissure. The reason we use this technique is that the design of this flap is simple, the pedicle will become the new commissure, and the transfer can be done in a single stage. To prevent oral stoma outcomes became too small, at least two-thirds of both lips must be preserved.

CONCLUSION

Noma is a rapidly progressive polymicrobial opportunistic infection resulting in orofacial gangrene. The etiopathogenesis of the disease has been attributed to extreme malnutrition, dehydration, and inadequate oral hygiene. This disease frequently leads to a mutilated face with severe functional impairments like trismus or ankylosis of the mandible, oral incontinence, also drink, eat and speech problems, and sequelae often result in social isolation. Reconstruction must be planned carefully to restore the natural contours of the lip, whereas the functional goals of lip reconstruction include maintenance of oral competence, sufficient oral access, and preservation of sensation. The Estlander flap is one of the treatment choices used for labial defects that involve the commissure.

CONFLICT OF INTEREST

There is no competing interest regarding the manuscript.

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AUTHOR CONTRIBUTION

All the authors are responsible for the study from the conceptual framework.

REFERENCES

1. Enwonwu CO. Noma-The Ulcer of Extreme Poverty. *N Engl J Med*. 2006;354:221-224.
2. Tonna JE, Lewin MR, Mensh B. A case and review of noma. *PLoS Negl Trop Dis*. 2010;4(12):e869.
3. Baratti-Mayer D, Gayet-Ageron A, Huggonet S, Francois P, Pittet-Cuenod B, Huyghe A, Pittet D. Risk factors for noma disease: a 6-year prospective matched case-control study in Niger. *Lancet Glob Health*. 2013;1:e87-96.
4. Enwonwu CO, Falkler WA, Philips RS. Noma (Cancrum Oris). *Lancet Infect Dis*. 2006;368:147-56.
5. Huyghe A, Francois P, Mombelli A, Tangomo M, Girard M, Baratti-Mayer D, et al. Microarray analysis of the microbiota of gingival lesions in noma patients. *PLoS Negl Trop Dis*. 2013;7(9):e2453.
6. Reddy BP, Reddy B S, Kiran G, Chembolu N. Cancrum oris: A devastating orofacial gangrene. *J NTR Univ Health Sci*. 2012;1:192-4.
7. Chidzonga MM, Mahomva L. Noma (cancrum oris) in human immunodeficiency virus infection and acquired immunodeficiency syndrome (HIV and AIDS): clinical experience in Zimbabwe. *J Oral Maxillofac Surg*. 2008;66:475-85.
8. Vaidya S, Agrawal A, Sharma VK. Cancrum Oris: A case report. *Indian J Otolaryngol Head Neck Surg*. 2006;58(4):411-413.
9. Baratti-Mayer D, Pittet B, Montandon D et al. Noma: an "infectious" disease of unknown etiology. *Lancet Infect Dis*. 2003;3:419-31.
10. Marck KW. A history of noma, the "Face of Poverty." *Plast Reconstr Surg*. 2003. 111:1702-7.
11. Montandon D, Lehmann C, Chami N. The surgical treatment of noma. *Plast Reconstr Surg* 1991;87:76-86.

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