ABSTRACT

Noma or cancrum oris was first described by Tourdes in 1848 as a ‘gangrenous affliction 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, destroying the soft parts and the bone and almost always quickly fatal…….’ This definition holds as true today as was in 1848. The only difference in clinical perspective is that the prognosis has improved. Noma comes from the Greek word ‘nomein’/ ‘nome’, meaning “to devour”. Greek historian Polybius (2nd Century before Christ) wrote ‘the wound made himself a nome’, meaning that the wound was becoming progressively larger.

Key words: Noma, cancrum oris, trismus, gangrenous, poor oral hygiene, Poverty and malnutrition

INTRODUCTION

When you hear its name for the first time, you don’t know what it is. When you hear its description, you can’t believe it. And when you see it with your own eyes, you’re never the same again. Noma or cancrum oris was first described by Tourdes in 1848 as a ‘gangrenous affliction 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, destroying the soft parts and the bone and almost always quickly fatal…….’ This definition holds as true today as was in 1848. The only difference in clinical perspective is that the prognosis has improved. Noma comes from the Greek word ‘nomein’/ ‘nome’, meaning “to devour”. Greek historian Polybius (2nd Century before Christ) wrote ‘the wound made himself a nome’, meaning that the wound was becoming progressively larger.(1) Noma is called the ‘face of poverty’.Poverty and malnutrition are directly related with the incidence of noma.
Noma is not caused by a specific pathogen but triggered by a consortium of micro organisms of which Fusobacterium necrophorum is a key component. Other pathogens include spirochaetes, Prevotella intermedia and Borellia vincenti. The organism are a part of normal oral flora, hence it implies that noma should be considered as an opportunistic infection.

Patients that survive the acute noma stages generally suffer from its sequelae, including serious facial disfigurement, trismus and ankylosis, oral incontinence, and speech problems. The complex facial defects can be classified with the NOITULP-system, as proposed by Marck in 1998. NOITULP is an acronym of Nose, Outer lining, Inner lining, trismus, upper lip, lower lip and particularities.
PATHOGENESIS OF TRISMUS IN NOMA:

<table>
<thead>
<tr>
<th>Process</th>
<th>Incidence</th>
<th>Effect</th>
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</thead>
<tbody>
<tr>
<td>scar formation, wound contracture and fibrosis in facial tissues (mucosa, facial muscles, skin)</td>
<td>always present</td>
<td>partial trismus up to fibrotic ankylosis</td>
</tr>
<tr>
<td>fibrosis and/or myostatic contracture of masticatory muscles</td>
<td>common</td>
<td>partial trismus</td>
</tr>
<tr>
<td>coronoid process hypertrophy causing impingement on zygoma</td>
<td>unknown</td>
<td>partial trismus up to ankylosis</td>
</tr>
<tr>
<td>formation of bony bridges between mandible and facial skeleton</td>
<td>common</td>
<td>extra-articular ankylosis</td>
</tr>
<tr>
<td>temporomandibular joint destruction</td>
<td>rare</td>
<td>decreased joint mobility up to intra-articular ankylosis</td>
</tr>
</tbody>
</table>

Gangrene -> scar formation, wound contracture and fibrosis -> TMJ destruction -> **Intra-articular ankylosis**
Pain, scar formation, wound contracture and fibrosis-> Myostatic contracture (ipsilateral or bilateral)-> **Hypertrophy of coronoid process** ->Partial trismus -> **Extra-articular fibrotic ankylosis**. If there is bony bridge formation -> **Extra-articular bony ankylosis**

**Case 1:**

- patient c/o inability to open mouth since age of 3 years.
- History of present illness – patient gives a history of suffering from an infection of the oral cavity when he was 2-3 yr old. The lesion was surgically excised and the defect covered with a distant flap following which there was restricted mouth opening progressing to the present condition. Relevant medical history; patient is anaemic with an Hb of 6 gm%.
- Family history - patient’s mother is hypertensive and diabetic.
- Past dental history: NRR
- Personal history: Brushes once daily, Bowel and bladder movements: not disturbed, sleep not disturbed.
General physical examination:

Moderately built and nourished, Pallor of the lower palpebral conjunctiva.
No signs of, icterus, cyanosis, clubbing and oedema. Vital signs are within normal range. Lymphnodes- non palpable.

Local examination:
Imaging:

OPG

MRI:
Preoperative work-up:

- All routine investigations
- Blood transfusion
- Nutritional support

Intubation: Fiberoptic intubation
Intraop:

Release of fibrotic band (intraorally)

Lateral cheek flap
Maxillo-mandibular fusion

Osteotomy to release the fusion
Mouth opening after the release 19 mm

Ipsilateral coronoidectomy
Excised coronoid

Mouth opening: 28mm
Contralateral coronoidectomy

Mouth opening: 45mm
Defect closure with collagen
Results (post op)

Case 2
Intraoperative steps

Post op results
Collagen:

- No need for second surgical site
- No contracture as seen in case of skin flaps
- Less bulk
- Most of the times flap is not a good option considering the medical status of the patient and the problems faced by the patients
- Cheap

Postoperative care:

- Immediate post op aggressive physiotherapy in form of active mouth opening exercises, joint ultrasound.
- Helps in preventing fibrosis of the surgical site by contracture
- one of the major factors that seems to influence the long-term result is postoperative physiotherapy.3

DISCUSSION

Noma (also referred to as cancrum oris, fusospirochetal gangrene, necrotizing ulcerative stomatitis, stomatitis gangrenosa)4 is a rapidly progressive, polymicrobial, often gangrenous infection of the mouth or genitals.

Fusobacterium necrophorum and Prevotella intermedia are thought to be key players in the process and interact with one or more other bacterial organisms (such as Borrelia vincentii, Porphyromonas gingivalis, Tannerella forsythia, Treponema denticola, Staphylococcus aureus, and nonhemolytic Streptococcus spp).5

The reported predisposing factors include:6,7

- malnutrition (particularly A-and B-vitamins) or dehydration
- poor hygiene, particularly oral
- unsafe drinking water
- proximity to unkempt livestock
- recent illness
- an immunodeficiency disease, including AIDS
Presentation

Noma (1836)
The mucous membranes of the mouth develop ulcers, and rapid, painless tissue degeneration ensues, which can degrade tissues of the bones in the face. In a condition sometimes called noma pudendi, noma can also cause tissue damage to the genitals.

Prognosis

The disease is associated with high morbidity and mortality and mainly affects children under the age of twelve in the poorest countries of Africa. Children in Asia and some countries of South America are also affected. Most children who get the disease are between the ages of two and six years old. The WHO estimates that 500,000 people are affected, and that 140,000 new cases are reported each year. The mortality rate is approximately 90 percent.

Treatment

Known in antiquity to such physicians as Hippocrates and Galen, noma was once reported around the world, including Europe and the United States. With improvements in hygiene and nutrition, noma has disappeared from industrialized countries since the 20th century, except during World War II when it was endemic to Auschwitz and Belsen concentration camps. The disease and treatments were studied by Berthold Epstein, a Czech physician and forced-labor prisoner who had recommended the study under Josef Mengele's direction.

The progression of the disease can be halted with the use of antibiotics and improved nutrition; however, its physical effects are permanent and may require oral and maxillofacial surgery or reconstructive plastic surgery to repair. Reconstruction is usually very challenging and should be delayed until full recovery (usually about one year following initial intervention). Children and other noma survivors in Africa are helped by a few international charitable organizations, such as Facing Africa, a UK registered charity that helps Ethiopian sufferers. There is one dedicated noma hospital in Nigeria, the Noma Children Hospital Sokoto, staffed by resident and visiting medical teams. In other countries, such as Ethiopia, international charities work in collaboration with the local health care system to provide complex reconstructive surgery which can give back facial functions such as eating, speaking and smiling. Teams of volunteer medics coming from abroad are often needed to support the local capacity to address the most severe cases, which can be extremely challenging even for senior maxillofacial surgeons. On 10 June 2010 the work of such volunteer surgeons was featured in a UK BBC Two documentary presented by Ben Fogle, Make Me a New Face: Hope for Africa's Hidden Children.
Conclusion

Noma robs many children of their future. The management of trismus in noma patients is a difficult surgical field. Trismus in noma is not caused due to dysfunction of mouth opening muscles. Their function is hindered by the influence of pathophysiological processes of the skin, mucosa and mouth closing muscles.

Physiotherapy should start after the first period of severe postoperative swelling. What is needed is not just the surgical trick; the art of medicine according to the rules of Hippocrates requires continuity.

‘Noma, should we care?’ yes, we should.

References

doi:10.1016/S0140-6736(06)69004-1. PMID 16829299.
[13]. Neville, Brad. Oral and Maxillofacial Pathology, 3rd Ed. Saunders Book Company, 062008. 5.11.2