Noma

Noma (Gr. numein: to devour) or cancrum oris is a terrible gangrenous disease which leads to severe soft and hard tissue destruction in the face (mouth, teeth, lips, nose, cheeks) with lasting disfigurement. It is associated with a high mortality.

The exact aetiology is not yet known, but it is thought that several factors contribute to this devastating illness. It is clearly a disease of poverty and social deprivation. Improvements in general socioeconomic status, public health and nutrition made that noma disappeared from all places except the most desperately poor and where severe malnutrition occurs. Several factors contribute, such as malnutrition with associated vitamin and trace element deficiencies, poor oral hygiene, a compromised immune status (malnutrition, measles, CMV infection, blood dyscrasia such as leukaemia), a lesion of the gingival mucosal barrier, a (bacterial?) trigger and inappropriate initial treatment. They probably act together to cause noma. Bacteria such as spirochaetes, *Prevotella intermedia* and *Fusobacterium necrophorans* are suspected to play a role in the acute pathology. However, it should be remembered that at present, most bacteria in the mouth cannot be cultured in vitro.

Although the disease existed in Europe and other parts of the globe, at present it is most common in Africa. The disease affects mostly children between 2-6 years but can occasionally appear in older children and even in debilitated adults (Auschwitz!).

The disease starts as an acute painful necrotising gingivitis (“trench mouth”), evolving to a necrotising stomatitis with ulceration and oedema of the cheek. The lesion tends to start at the alveolar margin in the premolar-molar region. It spreads very fast (1-2 days). Within a couple of days, a greyish area appears on the cheek. This becomes black and necrotic and has well defined margins. There is an offensive odour. The necrotic zone penetrates the cheek and has a typical cone shape (“cône gangréneux”). After the necrotic tissue has sloughed away, bone is exposed. Large bone sequesters may form, sometimes with destruction of maxilla and/or mandibule. It should be distinguished from pyogenic abscesses and Burkitt’s lymphoma. Secondary infection occurs rapidly, as can be expected.

Fever occurs in some patients. Many patients die due to starvation, septicaemia, or aspiration

pneumonia. Because of the high mortality in acute noma and the fact that it occurs in the poorest among us in areas with inadequate reporting, the burden of disease is difficult to determine for epidemiologists.

Noma, cancrum oris. Photo Cochabamba, Bolivia

Noma, cancrum oris. Face ulcer. Photo Cochabamba

The tissue defects are classified in 4 types:

1. Type I is the most common and consists of a localised cheek and commissural defect. It can be bilateral.
2. Type II includes the upper lip, and in some cases the nose and the palate.
3. Type III is located on the lower lip ± the mandible and floor of the mouth.
4. Type IV consists of major defects of the whole cheek, lips, palate, maxilla and can extend up to the orbit, eyelids and nose.

Treatment in the acute phase encompasses proper oral hygiene, mouth rinses with

chlorhexidine, antibiotics including penicillin and metronidazole against anaerobic bacteria, proper nutrition and vitamin/trace element supplements and treatment of any underlying medical conditions. The healing is characterized by ugly scars with fibrous tissue which tends to provoke strictures. After the acute phase, physiotherapy should be initiated to limit the strictures, fibrous scarring, trismus and to avoid bony ankylosis (bridging) between upper and lower jaw. Bundles of wooden spatulae in the mouth or more sophisticated devices (e.g. the Therabite) are used. At least a year after the initial disease, reconstructive craniomaxillofacial surgery for the sequelae can be considered. This should be done by experienced teams including specialised surgeons and anaesthesiologists (tracheostomies, fiberoptic intranasal intubation). Each case will require an individual approach.

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