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Mini-Review

Noma in Cambodia: scars from the past

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Background: Noma is an orofacial gangrene that tends to afflict starving and malnourished children. It has a high mortality rate, and even if the child survives, a lifelong deformity of the face occurs. There is a worldwide incidence of Noma in areas of mass poverty and famines, but it is rare in South East Asia. In Cambodia, the Children's Surgical Centre (CSC) has seen and treated 20 patients with facial deformity secondary to Noma occuring in the 1970s (during the "Pol Pot period").
Objective: A review and case report.

Keywords: Cambodia, Noma.

Noma is a rapidly spreading orofacial gangrene which mostly affects children between one and four years of age. It was recognized by the Greeks who named it, ("Noma" means "to devour"), and recorded by the Romans, but the first clear description came from Europe in 1595 [1]. By about 1880, it began to disappear from Europe with improvements in the standard of living when the population no longer suffered severely from famines and public health care improved. It still persists where poverty and famine are rife, and can reappear in unexpected sites such as in the Nazi concentration camps in Germany in the 1940s [2]. (For acute Noma, see **Fig. 1**).

In 1998, WHO estimated there were about 140,000 cases worldwide with a 79 % mortality [2]. The majority of these cases occur in Sub-Saharan Africa (the "Noma belt"), but Noma also occurs in Asia and South America. In 2003, the prevalence in Northwest Nigeria was said to be 6.4 per 1,000 children [3].

Causes of Noma

There are different theories regarding the causes of noma. In a setting of severe malnutrition and poor oral hygiene where there is a weakened immune system, an incidental systemic infection such as

Fig. 1 Progressive Noma.

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measles, malaria or typhoid can trigger a change in the microbial environment of the mouth which very quickly leads to an invasion of the gingivae ("acute necrotising gingivitis") and direct spread through the adjacent tissues to produce a localised area of gangrene [4]. There is dispute as to whether this infection is caused by normal saprophytic flora, by an invasion of a herpes virus, or by the invasion of fusiform bacteria and spirochaetes [1].

Most commonly, in starving and neglected areas of the world, the child dies, but if saved by adequate treatment with antibiotics, supplemental feeding and oral debridement, then characteristic defects are left around the mouth. This attempts to heal spontaneously, with scar contracture generally narrowing the mouth but leaving salivary incontinence, severe dental malposition and even maxillo-mandibular ankylosis [2, 4]. Even those few who survive Noma are disfigured for life, both functionally and cosmetically. They rarely receive adequate reconstructive surgery [2].

Treatment

The acute treatment of cases of noma is theoretically well defined: administration of antibiotics; debridement of oral lesions and maintenance of oral hygiene; correction of the nutritional deficits including the administration of vitamins and trace elements; and the prevention of contractures. However, in the usual context in which Noma occurs, medical attention is either not available at all or is so overwhelmed by the numbers of ill and starving people that adequate care cannot be given. Should the victim survive, then the defects can be classified into four types, depending on the sites of destruction, they are readily recognizable later in life unless treated by except reconstructive surgery. In Africa, there are hospitals specializing in the treatment of Noma patients, and some Universities in the developed world have taken an interest in this disease, and set up special programs to treat it (**Fig. 2**).

Methods

Patients who seemed to have sequelae of a disease resembling Noma were first seen at CSC shortly after the unit commenced operations in 1998. Several visiting Maxillo-facial and Plastic Surgery specialists from universities in the developed world subsequently saw these and other patients with similar deformities, and were all in agreement that such patients had suffered from classical Noma and had survived. All of the patients had one or more operations, usually by the visitors, not only in CSC but also in various other hospitals in Phnom Penh, and recently a search for the records of these cases was undertaken.

Results

Records of 20 distinct cases were discovered. For each of these, the records were retrieved and information and photos from them were compiled. Most of the patients came from rural areas and had been seen and treated several times over the past 8 years. Illustrative photos of the deformities are presented.

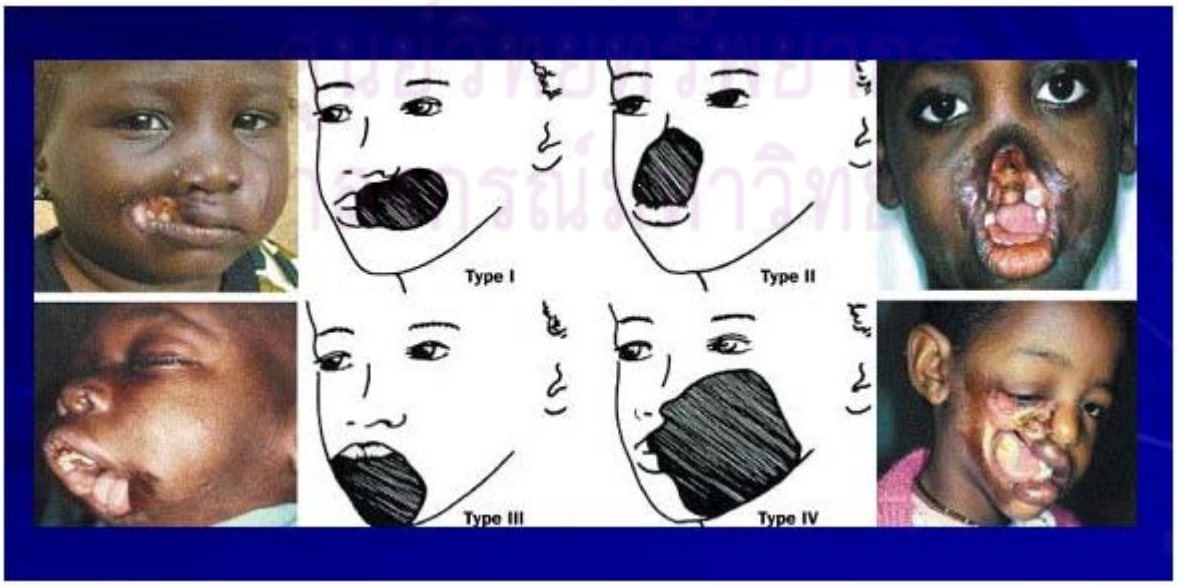

Fig. 2 Classification of Noma sequelae, with corresponding clinical cases [5].

Fig. 3 Representative cases of Noma sequelae as seen at Children's Surgical Centre (CSC) in Cambodia (1998-2006).

One of the patients, living not so far away, was asked to come for an interview, to provide a more precise history of the affliction and the reasons the patient had managed to survive. The interview took place in the consultation room at CSC. A staff member who spoke Khmer translated questions into Khmer for the interviewees, and then translated back their answers into English.

Interview summary

Miss Srey Sophea (not her name), a 27 year old female, came in with her mother for an interview at CSC on the 28th of September, 2006. She also gave us consent to publish her story and picture. Since Miss Sophea had been an infant during her infection with Noma, her mother answered most of the questions although she was vague about dates and ages.

Her mother stated that in 1978, when Sophea was born during the Pol Pot regime, she was generally able to eat twice a day and after birth did have enough milk to breast feed her baby. When asked, she said that she was of about the same weight when Sophea was a baby, 32 kg, as she is currently as a rather frail 59 years old lady.

Sophea was her third child. The first developed measles in 1977 as a baby, but healed uneventfully, and has survived to adulthood. The second died in 1977 or 78, when very young, of unknown cause but of normal weight at the time of death. Sophea caught measles when she was about 5 months old in 1978, and five days later, her mouth became infected and her lower lip turned black. She was treated with traditional medicine as nothing else was available. Her mother feared she would die, but continued to clean the wound around her mouth. Since the baby was unable to breast feed, her mother fed her by dropping milk into her mouth. Soon after the Pol Pot regime ended in January 1979 with the liberating Vietnamese troops, the baby was admitted to a hospital in Phnom Penh for 2 months. During this time, her mouth finally healed, and mother and the baby were able to return to her village when "the baby could already walk". Her mother says she never saw any other children with a condition like her daughter's.

Fig. 4 Miss Sophea in 2006, after reconstruction of the lower lip.

Miss Sophea first sought treatment for her damaged mouth in 2000, when she heard about cleft lip repairs at Children's Surgical Centre. She has now had 3 operations, and says she is happier than she was before, but has not yet married since none has asked her. She feels that she is still not pretty enough and would like her lower lip to be improved further.

Discussion

Noma is a debilitating and frequently fatal disease. What makes Noma even more tragic is that it is so relatively easy to prevent. Research shows that the single most highly correlated risk factor for Noma is severe malnutrition due to extreme poverty. The crippling and disfiguring injuries that result from Noma can be limited with early intervention, and the lifelong suffering caused to those few lucky enough to survive Noma can also be prevented, or at least aliviated. Unfortunately, conditions which provide fertile ground for Noma are also those which tend to prohibit timely and effective medical intervention.

Though Noma tends to be more common in developing countries, particularly those in sub-Saharan Africa, the incidence is not limited to that region. It seems that Cambodia may have had its own small Noma crisis. Most cases can be traced directly to the hardships suffered under the Pol Pot regime a few decades ago [6].

Cambodia

During the Pol Pot regime in the 1970s, Pol Pot and his party, the Khmer Rouge, sought to rebuild Cambodian society. The destruction of the current social order was necessary for this to occur. Education of any sort, and foreign education in particular, was frowned upon and among those selected for extermination by the Khmer Rouge were the educated classes, including doctors and other medical professionals.

Cities were evacuated, the population dispersed to the countryside, and normal life all but ground to a halt, resulting in famine. Additionally, the Khmer Rouge used extermination tactics similar to the Nazis, including organized starvation. Thus, Cambodia, in and around the time of the Pol Pot regime, was a fertile ground for Noma to rear its ugly head.

Of the 20 patients seen at Children's Surgical Centre (CSC) for treatment of damage resulting from prior Noma infection, the majority were of an age which shows that they were infants or very young children during these times and so particularly susceptible to Noma. All have subsequently been treated with various surgical techniques in order to relieve their condition, but the damage caused by Noma is so significant that complete repair is rarely if ever possible. These patients are living reminders of an era in Cambodian history that hopefully will never be repeated.

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Appendix

Fig. A1 A 3-year-old malnourished girl with acute orofacial Noma before removal of the tissue slough. The lesion has a well-demarcated perimeter surrounding a blackened necrotic centre (Courtesy from Noma Children Hospital, Sokoto, Nigeria) [7].

Fig. A2 A malnourished boy aged 22 years with a Noma lesion involving the orofacial tissues, lips, and nose, and extending to the infraorbital margin [7].