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Abstract. Oro-facial noma is an oral gangrene occurring in early childhood in extremely poor areas. As many as 70–90% of those with noma die, and to date, there is no satisfactory treatment to fight this disease. Within the context of the World Health Organization international program against noma, a 13-year retrospective study was carried out in Dakar, Senegal in an attempt to understand the epidemiology of noma. Between 1981 and 1993, 199 cases of noma were identified, among them; 36.7% were acute cases and 63.3% showed sequelae. Chronic sequelae of noma were seen in patients 2–41 years of age, but the acute phase of noma was found only in young children (77.7% in those 14 years of age, maximum age = 9 years, mean age ± SD age = 3.4 ± 1.9 years). A total of 73.1% of the cases with acute disease were reported in the Dakar, Diourbel and Kaolack regions during the dry season (57.0% of the cases). The lesions of progressive noma were localized mainly on the upper lip (42.4%) and the cheek (31.1%). A total of 96.9% of the patients with acute diseases were had poor general health with serious associated diseases; only 20.0% had a good vital prognosis. The development of epidemiologic surveillance programs for noma should be a public health priority in Senegal.

Oro-facial noma is a gangrenous disease found almost exclusively in young children in extremely poor socioeconomic living conditions. It develops very rapidly with serious associated diseases, such as protein deficiencies (kwashiorkor, marasmus), measles, and immune diseases. Oro-facial noma usually starts with an oral ulcero-necrotizing gingivitis.1 This gingival lesion evolves toward a facial edema that spreads from the inside to outside, rapidly destroys soft tissues and bones, and disfigures children horribly.1 The mortality rate of this disease is estimated to be between 70% and 90%,2 and surviving patients have serious functional and esthetic after-effects, for which there have not been any satisfactory therapeutic programs in developing tropical countries.2

In 1994, Dr. H. Nakajima (World Health Organization [WHO] Director-General) proposed the development of a five-step action program against noma.3 Epidemiological research is one of the main features of this program.2,3 The epidemiology of noma is poorly understood, making the development of primary and secondary prevention strategies difficult. No epidemiologic study has been published during the past two decades; data date back mainly to the period 1950–1965.3 Furthermore, there has not been any formal surveillance system set up for noma. However, a few cases of noma have been reported in the last 20 years in most developing countries, especially in Africa. The WHO has been expecting the emergence of a noma-affected zone in sub-Saharan countries, with a possible annual incidence estimated between 2 and 5 cases per 10,000 children 0–6 years of age.3 In 1994, within the context of the WHO international program against noma, a 13-year retrospective study was carried out based on cases seen at a national hospital in Dakar, Senegal. This was a joint project of the University of Dakar (Dakar, Senegal), the University of Lyon (Lyon, France), and the WHO.

MATERIALS AND METHODS

Senegal has an area of 196,772 km² and is divided into 10 regions with a population of 6,892,720 inhabitants (1993). Apart from densely populated areas around the capital Dakar and other large towns, the population is scattered sparsely in rural areas. The population median age is 18.0 years, the infant mortality rate is 8.6%, and the fertility synthetic rate is 6.4 children per woman. The main causes of death of children 0–5 years of age are diarrhea (23.8%), respiratory diseases (23.0%), and malaria (9.0%) (1993). The infantile population also suffer from malnutrition and dehydration, leading to marasmus and kwashiorkor.4

Senegal has a decentralized health policy based on primary health care practice.9 The Senegalese health care network includes 17 hospitals, 51 health centers, 639 health posts, 10 districts for serious endemic diseases, 10 regional health teams, and 37 centers for social advancement and rehabilitation. Until 1994, the National Hospital Center of Dakar was the reference site for the treatment of maxillofacial diseases. Traditional medicine, which operates without the supervision and control of the Senegalese Ministry of Health and Social Services, also plays a significant but unevaluated part in the health care system.

This retrospective study, which sought to identify all confirmed cases of oro-facial noma, was based on 1981–1993 data in the clinical records of the National Hospital Le Danlec (Dakar, Senegal). The records were compiled by a local maxillofacial physician (BD). The clinical data were summarized in WHO case report forms (CRFs). Each CRF was reviewed by a staff epidemiologist (DMB) in association with the physician for the interpretation and determination of patient status and the detection of discrepancies.

Oro-facial nomas were classified in progressive or residual stages that included the recording of functional lesions, such as permanent constriction of the jaw (ankylosis). Localization of lesions was based on the WHO classification,10 which distinguishes commissure, cheek, lip, and complex lesions. The sociodemographic data included age, sex, geographic location, as well as year of consultation and clinical information, such as type and stage of lesion, prognosis, clinical symptoms, and general health. The geographic regions were grouped according to standard census divisions.9
For each referred case, this descriptive statistical analysis distinguished noma sequelae, for which the initial stage of development cannot be evaluated clinically, from the acute phase of the disease, for which the extremely rapid clinical evolution (a few weeks) reflects the incidence of the disease. For the latter, the incidence per million was defined as the total number of cases per year of exposure, which was determined by summing census figures for each group at risk.

RESULTS

From January 1, 1981 to December 31, 1993, 199 cases of noma were identified through the CRFs. They were reported from 8 regions of Senegal, among which Dakar (31.9%), Diourbel (25.8%), and Kaolack (14.7%) reported 72.4% of the total number of cases. There were 95 (47.7%) male and 104 (52.3%) female patients.

Chronic sequelae of oro-facial noma. A total of 63.3% of the cases showed chronic sequelae of acute disease. The age of the patients ranged from 2 to 41 years, with a median age of 18.0 years and a mean ± SD age of 18.3 ± 7.3. Thirteen patients (10.3%) were less than 10 years of age and 35 patients (27.7%) were more than 22 years of age. There were 55 (44.0%) male and 74 (56.0%) female patients with a mean ± SD age higher for females (4.0 ± 2.0 versus 2.8 ± 1.8 years; P < 0.05). These cases of noma were reported mostly from Dakar (30.2%), Diourbel (27.0%), and Kaolack (14.0%).

Most of the lesions were localized on the upper lip (37.3%) and the cheek (31.9%), whereas less were seen on the lower lip and commissure lesions (11.9% and 10.3%, respectively). The majority were highly widespread and severely disfiguring complex lesions.

Acute phase of oro-facial noma. Seventy-three cases were in the acute phase of the disease. All were young children. No lesions were found in children less than 1 year of age and in those more than 9 years of age. The mean ± SD age of the patients was 3.4 ± 1.9 years (median = 3.0 years, mode = 2.7). A total of 77.7% were between 1 and 4 years of age and 40.2% were less than 3 years of age; the maximum age was 9 years. The frequency of the patients (54.1% versus 45.9%; P < 0.05) and the mean age of patients (2.8 ± 1.8 versus 4.0 ± 2.0 years; P < 0.05) with acute cancrum oris differed between males and females. Cases of noma were reported from 8 regions; most (73.1%) were from Dakar (34.2%), Diourbel (23.6%), and Kaolack (15.3%).

The annual incidence of progressive noma in children 1–4 years of age was 4.2 cases per million. Figure 1 shows the highest incidence of acute oro-facial noma per region for the children 0–4 years of age. The annual incidence for children 5–9 years of age was 1.2. The average monthly rate of diagnosis for acute noma was 4.0. The highest numbers of cases were reported in March (14) and November (9), and the lowest were reported April and July (2 and 3, respectively); 57.5% of the cases were reported between November 1 and March 31. However, sporadic cases were reported throughout the year. The number of cases per year of consultation reported in 1989 and 1990 was 12 and 11, respectively, and the median annual number was 6.0 cases.

A total of 42.4% of the lesions for which patients with acute cancrum oris sought medical treatment were localized on the cheek, 31.1% were on the upper lip, 9.1% were on the lower lip and the commissure, and 7.6% were on various parts of the face. A total of 96.9% of the children with progressive oro-facial noma had poor general health. The most frequently named associated diseases were anemia (48.0%), dehydration (18.0%), undernutrition (14.0%), measles (4.0%), and diarrhea (2.0%). Based on the medical evaluation, 20.0% had a good vital prognosis (stabilization of the lesions) without any consideration of residual functional and/or-esthetic effect, 10.0% died, 6.0% were in a critical situation, and 3.3% could not expect any health improvement.

The other 60.0% were not in the hospital system, indicating that they did not receive any treatment. As a result, those children had an extremely unfavourable prognosis.

DISCUSSION

The epidemiologic data in the 328 publications referenced on oro-facial noma at WHO since the beginning of the century vary greatly. They do not include any current evaluation of the annual incidence of oro-facial noma in Africa. In 1966, Tempest analyzed 250 cases over a 2-year period (1963–1965). Enwonwu studied 69 cases in Nigeria during the period of 1963–1965 and Reynaud and others examined 47 cases in Senegal in 1965. In 1990, Oosuji analyzed 58 cases of necrotizing ulcerative gingivitis and 5 cases of cancrum oris at the Ibadan Hospital in Nigeria. Adolph and others in 1996 described the experience at the Galmi Hospital in Niger, with 50 operative patients of a group of 300 referred cases. Since 1991, WHO has recorded cases of noma in 23 countries. Therefore, our retrospective study is the first to investigate a long and recent period of time and a large number of cases.

In the absence of a representative sample, WHO recommends a method used in our study for calculating the estimate of the annual incidence of noma based on the following considerations: 1) the only data accessible was the number of cases of noma referred to treatment centers during a specific period; 2) the number of cases referred to and reaching the treatment centers (R) constitute a certain percentage (X) of total surviving cases (S); 2) the number of cases referred to treatment centers (R) constitute a certain percentage (X) of total surviving cases (S) with S = (R × 100)/X; and 3) the number of surviving cases is a percentage of total incidence that is dependent on the case survival rate (y/100). Therefore, the total incidence (I) is estimated by I = (S × 100)/y. The percentage of referred cases in relation to all survivors has to be taken into account. A total of 63.3% of the reported cases in the study were chronic. It can suggest that the reported cases that are chronic are an over-representation of the chronic survivors of acute disease. Within the context of a Delphi-type consensus conference, WHO has evaluated the percentage of referred cases for the African continent in a 5–20% bracket. The increasing number of wars in the Third World and the epidemic of acquired immunodeficiency syndrome (AIDS) have increased the likelihood that the number of cases of noma will increase, particularly since the incidence of this disease has been largely underestimated for many years. Because of the specific constraints in Senegal, which were identified through a WHO mission in 1994, a low bracket of referred cases tends to be accepted. The main constraints that are unfavorable to an effective treatment against noma are the size of the country.
with a difficult means of communication, the pyramid-shaped organization of the health care system, the population superstition toward noma, and the importance of traditional medicine.\textsuperscript{15}

The mortality rate of the patients that were not treated was approximately 90%. Death is rarely due to local destruction and results from predisposing illnesses and complications, such as aspiration pneumonia, intractable diarrhea, or resultant severe dehydration.\textsuperscript{5} Therefore, the annual incidence for the Senegalese population at risk would be included between 2.8 and 8.4 cases per 10,000 children 1–4 years of age, that is, between 252 and 755 new cases per year. The unpublished study cited by WHO\textsuperscript{6} of the cases referred in Niger in 1991–1992 reported concluded an annual incidence between 7 and 14 cases per 10,000 children 0–6 years of age with the same method.\textsuperscript{3} In some African areas, WHO has reported a possible incidence between 2 and 4 cases per 10,000 children.\textsuperscript{3}

The risk factors of the incidence of progressive noma can be addressed within the specific context of the referred cases. The epidemiologic features described in this study, such as higher incidence in children, associated diseases, and seasonality, are consistent with previous studies.\textsuperscript{2,11,16} As for the sex ratio, it was discussed and highly debated in the literature of the 1960s. There is currently no scientific evidence showing that the sex ratio has to be considered as a representative risk factor and this is not an indication of a gender difference in the presentation or development of this disease. Additional work should consider differences in behavior in relation to resorting to care. These differences were identified in most African sociologic and behavioral studies\textsuperscript{17} and cited in the WHO Senegal report on noma.\textsuperscript{15} Since male
children are preferentially given medical care, this might account for the young age bias for males.

Our results confirm that acute noma is a specific disease that occurs in early childhood. The literature from 1955 to 1965 reported a peak incidence of this disease in children 2–5 years of age.2 In 1997, WHO analyzed a consensus from a larger age bracket, that is, children 0–6 years of age.6 However, a few typical cases were recently reported in adults whose main risk factor seemed to be immunodeficiency.1 Our results are consistent with these estimations, which identified a group at risk 1–5 years of age, 40.3% of the sick children being 1–2 years of age, no sick child less than 1 year of age. This childhood period correlates well with the eruption of primary teeth, which was described as being associated with gingival inflammatory modifications likely to lead to an ulcero-necrotizing gingivitis on deficient grounds (i.e., human immunodeficiency virus infection, measles, childhood diseases).15

These children have serious associated diseases. At the time of consultation, 96.9% of the patients were in poor general health. There is a consensus in the literature that protein malnutrition, secondary infection, and vitamin and mineral deficiencies play a part in the predisposition to the development of noma.2,7,17 Reports of 4 adults and 5 children with AIDS and positive human immunodeficiency virus serology who developed noma were recently published.18,19 A relationship between protein nutritional content and the likelihood of developing oro-facial noma is a plausible assumption.17,20 Senegalese mothers and infant health services have reported that breast-feeding lasts until a mean age of 2 years.21 The weaning period, during which the child switches from a rich diet to a cereal soup, lasts an average of 19 months.21 The child is completely weaned from the age of 3.5 years, which corresponds in our study to the mean age of the patients in the acute phase of noma.

Seasonality is a factor that should be considered in future studies; 57.0% of the patients consult health centers from November to March, which corresponds to the dry season in tropical countries. This period has been described by some researchers as hungry months.4 In Senegal, it is characterized by an upsurge of endemic sources of malaria, measles, dysentery, and intestinal parasites.8

Three regions of Senegal (Dakar, Diourbel, and Kaolack) accounted for 72.4% of the cases of noma. This may reflect the influence of environmental factors although this point should be verified by a comparison of noma with the reporting of other, unrelated diseases such as Buruli ulcer. The current migration pattern is from rural areas to the capital of Dakar, and is leading to fast and uncontrolled urbanization with the formation of squatter camps, and all the consequences of insalubrity, poverty, and malnutrition. The Kaolack region is one of the main groundnut-oil producers of the African continent. Its inhabitants have a totally unbalanced diet with scarcely any protein. The Diourbel region has a very hot pre-desert climate. However, when observing our results, one has to take account that a low number of consultations in other regions does not necessarily indicate the absence of cases. The geographic barriers for children from northern and southern regions of Senegal in getting to the hospital in Dakar are evident. These children do not have the time or materials to realize a long journey in the difficult conditions that require many days of travel.

In conclusion, the development of epidemiologic surveillance programs for noma seems to be a public health priority. More precise data from chart reviews or prospective series are needed. It is important that the duration of noma applies only to the necrotizing processes that begin in the mouth and show the characteristic features and course.1 A routine program should be developed in primary health centers to identify the areas that are more likely to have outbreaks. Furthermore, other explanations should be explored, particularly in etiologic research.22 The estimated incidence of the reported cases and an extremely unfavorable prognosis, when hospital treatment leads to functional, esthetic, and social after-effects, are factors in favor of the development of a WHO program against noma to detect and treat cases early at the initial stage of the disease, before the development of gangrene.

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