Am J Med Genet 1986, 23: 869-901.

7. Ohno T, Tsuchida H, Fukuhara N, *et al.* Adrenoleukodystrophy: A clinical variant presenting as olivo pontocerebellar atrophy. J Neurol 1984,231:167
8. Moser HW, Moser AE, Trojak JE, *et al.* Identification of female carriers of adrenoleukodystrophy. J Pediatr 1983, 103: 54.
9. Rizzo WB, Leshner RT, Odone A, *et al.* Dietary erucic acid therapy for x-linked adrenoleukodystrophy. Neurology 1989, 39:1415-1422.
10. Auborg P, Blanche S, Janbaque I, *et al.* Reversal of early neurologic and neuroradiologic manifestations of x-linked adrenoleukodystrophy by bone marrow transplantation. N Engl J Med 1990, 322:1860-1866.

Noma Neonatorum

N.C. Prajapati
P. Chaturvedi
R.R. Bhowate
S. Mishra

Noma, a Greek word meaning to *'de vour'* was described by Tourdes in 1848 as reported by Tempset(l). The term noma neonatorum (NN) was coined by Ghosal *et al.* in 1977(2). It is a rare gangrenous disease, that results in mutilating loss of tissue. We report two such cases along with review of literature.

From the Neonatal Unit, Departments of Pediatrics and Dental Surgery, Mahatma Gandhi Institute of Medical Sciences, Sevagram, Wardha 442 102.

Reprint requests: Dr. N.C. Prajapati, Reader in Pediatrics and Incharge Neonatal Unit, M.G. Institute of Medical Sciences, Sevagram, Wardha 442 102.

Received for publication: August 3,1994; Accepted: November 4,1994

Case Reports

Case 1: A male baby, first of twins, was delivered vaginally at 34-36 weeks of gestation with a birth weight of 1340 g and Apgar score of 9, 10 and 10 at 1, 5 and 10 minutes, respectively to a second gravida. The second male twin weighed 1240 g.% Both of them were kept in Special Care Neonatal Unit (SCNU) and fed expressed breastmilk (EBM) by nasogastric (NG) tube.

On 8th postnatal day, the first of the twins developed induration of lips and erythema of labial, palatal and alveolar mucosa with few black spots on the gum pad. Intravenous benzyl penicillin, gentamicin and metronidazole were started after taking samples for culture. By 10th day, there was conjunctivitis of both eyes and vesicular necrotic lesions developed in the groin. In view of clinical deterioration, nasogastric feeds were discontinued and cefotaxime was added to the therapy. Sclerema along with erythema and edema of the scrotum were also noticed. Culture reports (taken

from several sites) revealed a polymicrobial growth, with *Pseudomonas aeruginosa* being the most common organism from all sites and was sensitive only to ciprofloxacin and amikacin. Antibiotics were, therefore, changed to ciprofloxacin and amikacin. An antibiotic—antifungal solution was applied to local lesions every two hourly. This antibiotic antifungal solution was prepared fresh everyday, by adding 20 mg of intravenous preparation of ciprofloxacin, 20 mg of IV preparation of metronidazole and 10,000 IU of nystatin suspension to 500 ml of distilled water. A remarkable improvement was noticed and by 15th day NG feeds were resumed. Antibiotics were given for three weeks. Necrotic areas shed by 38th day leaving partial loss of upper and lower lips, upper gums, anterior palate and tip of the tongue (*Fig. 1).* The second of the twin and mother were normal.

Case 2: A full term female baby was delivered to a primi mother by Cesarean section. The birth weight was 2220 g and Apgar score 2, 4 and 8 at 1, 5 and 10 minutes, respectively. She was being managed in the SCNU for HIE grade II and received IV benzyl penicillin, gentamicin and phenobarbitone. Blood cultures sent before starting antibiotics were sterile. On day 7 she developed erythema and induration of both lips with blackish discoloration. Based on our experience with the earlier case of noma (Case 2), same treatment, *viz.,* IV ciprofloxacin, amikacin, metronidazole and cleaning of lesions by antibiotic antifungal solution was started. By the 10th day, the baby improved, lesions started regressing and nasogastric feeding with EBM was started. By day 16 the lesions had healed completely and breastfeeding was resumed. Antibiotics were continued for three weeks. There was no scar or mutilating loss of tissue.

Discussion

Noma and noma neonatorum are considered different entities. The former has been described in children aged 2-5 years and is caused by *Borellia vincentie, Bacillus fusiformis, Bacteroids* and *Staphylococcus* species, while the latter is seen within the first month of life and *Pseudomonas aeruginosa* is the most common causative organism(3,4).

On review of the available literature, only 53 cases of NN have been reported. Noma in newborns was almost unknown till 1977, when Ghosal *et al.* from Calcutta reported 48 cases and coined

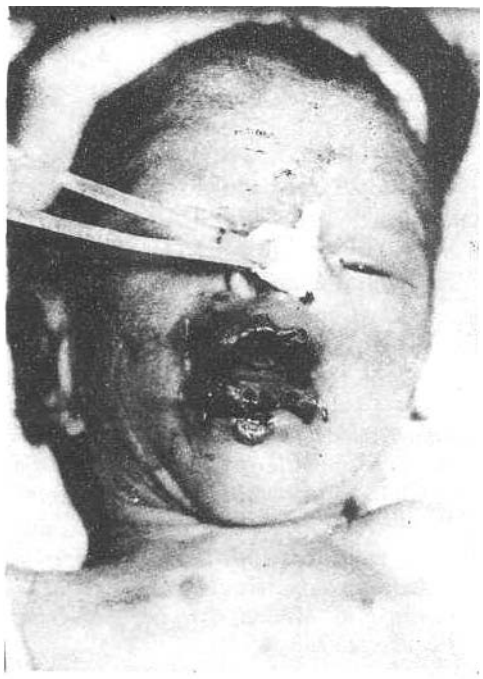

Fig. 1. Case 1 with mutilating loss of oral tissue

this entity as noma neonatorum(2). However, one case reported in 1930 is probably the first case of noma in a neonate(l). The other reported cases of NN are, one each from Washington(3), Israel(5), China(6) and India(7).

Noma neonatorum has no predilection for birthweight or gestation. Oral cavity is the most common site of involvement, the others being nose, eyelid, umbilicus, scrotum and groin. Onset of the disease has been reported to range from 3rd postnatal day to 120 days. *Pseudomonas aeruginosa* has consistently been isolated from all cases. The disease is fatal in majority of the cases with only 6 survivors amongst 54 cases (including two of ours) reported till date. Mutilating loss of tissue has occurred in all patients except in Case 2 of the present report.

Noma neonatorum needs to be treated aggressively with antibiotics cover against *Pseudomonas* species. Apart from supportive care, local measures like, repeated irrigation of the local lesions seems beneficial as the pooled secretions have a heavy concentration of causative organisms and the concentration of antibiotics in the secretions attained by systemic administration is doubtful. Extensive surgical debridgement is contraindicated in these cases and reconstructive surgery is advocated at least one year after the resolution(8,9).

REFERENCES

1. Tempest MN. Cancrum oris. Br J Surg 1966, 53: 949-969.
2. Ghosal SP, Chaudhari M, Dutta N, Sarkar AK, Mukherjee AK, Sen Gupta PC. Noma Neonatorum. Indian Pediatr 1977,14: 709-714.
3. Eisele DW, Inglis AF Jr, Richardson MA. Noma and noma neonatorum. Ear Nose Throat J 1990, 69:119-123.
4. Ghosal SP, Sen Gupta PC, Mukherjee AK, Choudhury M, Datta N, Sarkar AK. Noma neonatorum: It's etiopathogenesis. Lancet 1978, 2: 289-290.
5. Alkalay A, Mogilner BM, Nissim F, Barak Y, Handzel ZT, Ostfeid E. Noma in full term neonate. Clin Pediatr (Phila) 1985, 24: 528-530.
6. Lin JY, Wang DW, Peng CT, Tsai FJ, Chiou YM, Tsai CH. Noma neonatorum: An unusual case of noma involving a full term neonate. Acta Pediatr 1992, 81: 720-722.
7. Borle RM, Agarwal M. Noma neonatorum. Int J Oral Maxillofac Surg 1987,16: 626-629.
8. Griffin JM, Back DE, Nespec JA, *et al.* Noma-Report of two cases. Oral Surg 1983,56: 605-607.
9. Adekeye EO, Ord RA. Cancrum Oris: Principles of management of reconstructive surgery. J Maxillofac Surg 1983,11:160-170.