# *PSEUDOMONAS* SEPSIS WITH NOMA: AN ASSOCIATION?

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## *ABSTRACT*

***We report here a 2.5-year-old male child with community-acquired Pseudomonal sepsis showing the characteristic lesions of ecthyma gangrenosum. The child had development of gangrenous changes of the nose and face – the 'cancrum oris' or 'Noma'. We highlight the possible association of Pseudomonas sepsis and Noma, with malnutrition playing a central role in causing both the diseases.***


***KEY WORDS:*** **immunodeficiency; infection; malnutrition; poverty**

## INTRODUCTION

*Pseudomonas* usually infect surfaces of wounds or burns.[1] *Pseudomonas* septicemia occurring in normal children is a rare event.[1] Noma or cancrum oris is a devastating childhood infectious disease, which destroys the soft and hard tissues of the oral and para-oral structures by causing oro-facial gangrenous lesions.[2],[3] We report here a case of Noma in child with *Pseudomonal* sepsis. Such an association has not yet been reported in older children.

## CASE REPORT

A 2.5-year-old, unimmunized male child presented with high-grade fever for 3 weeks, cough and vomiting for 4 days, and irritability for 1 day. He had chronic right-sided otorrhea.

On examination, he had normal vital parameters with Grade-III protein energy malnutrition by the Indian Academy of Pediatrics (IAP) classification (weight – 9 kg, <5th percentile, expected of 13.6 kg). His height was 84 cm (<5th percentile) for an expected value of 93 cm. Pallor and platynychia were noted. Bilateral purulent rhinorrhea with erythema of nasal septum and turbinates was present. Right-sided otorrhea and clinical features of rickets were seen. Respiratory examination revealed bilateral crepitations. He had firm hepatomegaly (5 cm with span of 8.5 cm) and splenomegaly (2.5 cm). Meningeal signs and focal neurological deficits were absent. After 36 h of admission, he developed rapidly progressive gangrenous patch on the nose with surrounding cellulitis ('Noma'). Right-facial paralysis was noted. Erythematous papular lesions (varying from size of petechiae to 2-cm diameter) were noticed on the trunk and lower limbs, some of which formed blisters, that burst to leave behind typical lesions of 'ecthyma gangrenosum' over the next few days. The nasal gangrene also sloughed over 1 week.

Investigations revealed leukocytosis (cell count of 49 000/cumm), raised ESR (44 mm) and normal liver and renal function tests. Blood culture showed growth of *Pseudomonas aeruginosa*, as also the nasal swab and the wound swab from the lower limbs. *Pseudomonas* isolated from the blood culture was susceptible to ceftazidime, piperacillin, gentamycin, amikacin, and ciprofloxacin. The nasal swab and wound swab on the lower limbs also showed growth of *Pseudomonas*, which was susceptible to ceftazidime, amikacin, and ciprofloxin. Serological tests for syphilis and hepatitis B were negative. ELISA test for HIV – antibody was negative. Serum immunoglobulin levels were normal. Chest radiograph revealed bilateral bronchopneumonia. CT scan of the brain and temporal bone was normal except for maxillary sinusitis. Nasal septal biopsy showed panninculitis. The patient was treated with a 4-week course of intravenous ceftazidime, amikacin, and vancomycin. He recovered with persistence of the right facial palsy. Plastic surgical nasal reconstruction was advised at discharge.

## DISCUSSION

Usually Pseudomonal infections are seen in patients with burns, cystic fibrosis, malignancies, prematurity, immuno-deficiencies, HIV infection, immuno-suppressive therapy, or in malnourished children.[1],[4] *Pseudomonas* elaborates various products like lecithinase, collagenase, lipase, and hemolysins, which are proteolytic and may be causative in localized skin necrosis.[1] The characteristic skin lesion is that of 'ecthyma gangrenosum,' which starts as pink macules, progressing to small cutaneous hemorrhagic nodules and eventually causing necrotic areas with eschar formation surrounded by an intense red areola.[1],[4] In addition to skin infection and septicemia, meningitis, pneumonia and urinary tract infections can be caused by *Pseudomonas*.[1] *Pseudomonas* septicemia occurs more frequently in children with indwelling intravenous or urinary catheters.[1] Community – acquired *P. aeruginosa bacteremia* and sepsis has been occasionally reported in healthy infants.[5] Our patient had such a community-acquired *Pseudomonal* infection. He was severely malnourished, which was probably the underlying cause of such severe *Pseudomonal* sepsis (in the absence of other immunodeficiencies).

'Noma' has been known to mankind since its first report by Hippocrates in the fifth century BC with an increase in the understanding of the disease between the 16th and 19th century.[3],[6] The risk factors for Noma include poverty, malnutrition, immunosuppression (including HIV infection), poor oral hygiene, unsanitary environment, leukemia, and infectious diseases caused by measles and herpesviridae.[2],[3],[6],[7] It is usually seen in children between the ages of 3 and 12 years mainly in the developing countries especially sub-Saharan Africa.[3],[6] WHO (1998) estimated a worldwide incidence of 1 40 000 cases per

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year with prevalence of 7 70 000 people living with Noma sequelae (in 1997).[3] *Fusobacterium necrophorum* and *Prevotella intermedia* are the suspected causative organisms.[2],[6],[8] Other reported organisms isolated from the Noma lesions include alpha-hemolytic *Streptococci*, *Actinomyces* spp., *Peptostreptococcus micros*, *Veillonella parvula*, *Staphylococcus aureus*, *Corynebacterium pyogenes*, *Bacteroides fragilis*, *Bacillus cereus*, and *Pseudomonas* spp.[3],[8] Children at risk for Noma have been seen to have low plasma concentrations of zinc, retinol, ascorbate, and essential amino acids with increased plasma and saliva levels of free cortisol.[6] These nutritional deficiencies with infections like measles or herpesviruses, may cause impaired oral mucosal immunity.[6] Enwonwu et al. postulate that evolution of the oral mucosal ulcers including acute nercotizing gingvitis to Noma may be triggered by a consortium of microorganisms including *Fusobacterium microphorum* (which can elaborate dermonecrotic toxic metabolites and is acquired via fecal contamination resulting from shared residential facilities with animals and poor environmental sanitation).[6] The mortality rate in inappropriately treated or untreated patients with Noma approaches 70–90%.[2],[6] The sequelae include stricture of mouth, severe dental malposition, salivary incontinence, and loss of maxilla (with resultant defective speech and nasal regurgitation).[3] The oro-facial destruction necessitates plastic surgical reconstruction.[2]

'Noma neonatorum,' a distinct entity, is characterized by gangrenous process of the nose, oral cavity, eyelids, and perineum, and has been seen in cases with *Pseudomonas* sepsis in premature infants.[3],[9] In fact, Freeman et al. have questioned the distinction between 'Noma neonatorum' and neonatal presentation of 'ecthyma gangrenosum.'[9] The infectious organism in Noma neonatorum is usually *Pseudomonas* and is seen in neonates with *Pseudomonal* sepsis, the disorder being fatal in almost all cases (because of irreversible septicemia).[3],[10] According to Baratti-Mayer et al., Noma neonatorum is a discrete pathological entity and should not be confused with the classic form of Noma.[3] Although older children with *Pseudomonal* sepsis and Noma have not been reported so far, this association is likely. The child in this study had severe malnutrition, which can predispose to both, *Pseudomonal* infection and Noma. Hence, we have reported this case to increase the awareness of the medical fraternity about the possibility of their co-existence.

## REFERENCES


1. Feigin RD. Pseudomonas infections. *In:* Feigin RD, Cherry JD (eds). Textbook of Pediatric Infectious diseases, Vol. I. Philadelphia: W. B. Saunders Company; 1981. p. 948-52.
2. Enwonwu CO, Falkler WA, Idigbe EO. Oro-facial gangrene (noma/cancrum oris): pathognetic mechanisms. Crit Rev Oral Biol Med 2000;11: 159-71.
3. Baratti-Mayer D, Pittet B, Montandon D, Bolivar I, Bornand JE, Hugonnet S, Jaquinet A, Schrenzel J, Pittet D. Noma: an "infectious" disease of unknown aetiology. Lancet Infect Dis 2003;3:419-31.
4. Ng W, Tan CL, Yeow V, Yeo M, Teo SH. Ecthyma gangrenosum in a patient with hypogammaglobulinemia. J Infect 1998;36:331-5.
5. Wu BY, Peng CT, Tsai CH, Chiu HH. Community-acquired Pseudomonas aeruginosa bacteremia and sepsis in previously healthy infants. Acta Paediatr Taiwan 1999;40:233-6.
6. Enwonwu CO, Falkler WA Jr, Idigbe EO, Afolabi BM, Ibrahim M, Onwujekwe D, Savage O, Meeks VI. Pathogenesis of cancrum oris (noma): confounding interactions of malnutrition with infection. Am J Trop Med Hyg 1999;60:223-32.
7. Lazarus D, Hudson DA. Cancrum oris — a 35-year retrospective study. S Afr Med J 1997;87:1379-82.
8. Falkler WA Jr, Enwonwu CO, Idigbe EO. Microbiological understandings and mysteries of noma (cancrum oris). Oral Dis 1999;5:150-5.
9. Freeman AF, Mancini AJ, Yogev R. Is noma neonatorum a presentation of ecthyma gangrenosum in the newborn? Pediatr Infect Dis J 2002;21:83-5.
10. Eisele DW, Inglis AF Jr, Richardson MA. Noma and noma neonatorum. Ear Nose Throat J 1990; 69:119-20, 122-3.






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