

Noma-like lesion in a patient with acute promyelocytic leukemia

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The term noma is used to describe a spreading, invasive gangrene with edema of the face which starts as an ulcer of the mucous membrane and extends from in outwardly, rapidly perforating and destroying soft tissues and bone and almost always rapidly fatal.^(1,2) The disease occurs mainly in children from deprived areas with low levels of hygiene. Cases of children are common and occur mainly in under-developed countries; adult cases are rare and are reported predominantly in developed countries. However, some adult cases can be attributed to malnutrition and to a predisposition to infectious disease.

Leukemia is increasingly being recognized as an associated factor.⁽²⁾ Patients with leukemia are at a high risk of developing nomas during chemotherapy, as this can induce agranulocytosis.⁽³⁾ Regardless of the type of initial lesions, gangrene always develops rapidly, but remains well defined.⁽³⁾ Most patients do not consult their doctor until the disease is at an advanced stage, its onset and progression remains a mystery.⁽¹⁾ The prognosis for childhood noma has improved dramatically since the discovery of antibiotics, prior to which there was only a 15% survival rate whereas now only 15% succumb to it.⁽²⁾ Prompt and appropriate antibiotic therapy can ensure the patient's survival with an aesthetically acceptable outcome.⁽³⁾

A 17-year-old Caucasian male, who had had throat pain and odynophagia for one week, was submitted for blood tests (Hemoglobin: 7.0 g/dL; Hematocrit: 21%; WBC: 33×10^9 cells/L and platelets: 38×10^9 cells/L). Acute promyelocytic leukemia (M3) was diagnosed and the patient was treated with daunorubicin plus

vesanoid. A physical examination demonstrated an extensive necrotic lesion in the M. palatoglossus, soft palate, uvula and right tonsil, accompanied by an unpleasant odor (Figure 1). Samples were collected for biopsy and culture. The histopathologic findings showed non-specific chronic diffuse inflammation infiltrated by necrotic areas and numerous bacteria. The culture identified *Enterococcus spp*, *Staphylococcus aureus* and *Candida SP*. In accordance with the histological and bacteriological findings it was concluded that this was a noma-like lesion. The patient then received antibiotic therapy using ceftazidime, amikacin, vancomycin, fluconazole and penicillin G. The patient developed pneumonia and sepsis and was treated in the intensive care unit. Total recovery albeit with extensive loss of soft palate tissue was attained in 45 days.

Cancrum oris (noma) occurs predominantly among deprived and poorly nourished children in developing countries, who have poor oral hygiene. It is often associated with infections such as measles and rubella.^(1,3) In 90% of cases, nomas develop before the age of 10 years old.⁽⁴⁾ Other commonly associated diseases include: typhoid fever, whooping cough, typhus, syphilis, tuberculosis and leukemia in descending order of frequency.^(1,3,4) To the best of our knowledge, there are only five cases reported concerning nomas or noma-like lesions in patients with oncohematological malignancies in PubMed (Table 1).

It is difficult to pinpoint any specific triggering agent in the complex microbiota of a noma.⁽¹⁾ *Streptococcus*, *Fusobacterium* and *Bacterioides* have been associated with cancrum

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Figure 1 - Extensive necrotic lesion in the m. palatoglossus, soft palate, uvula and right tonsil with foul odor

Table 1 - Case reports published in PubMed about Noma and Noma-like lesions in patients with oncohematological malignancies

Case	Author	Year	Disease	Age/gender of patient	Debilitation	Microbial agents
1	Weinstein et al. ⁽⁵⁾	1974	Cancrum oris-like	27/F	Acute myelogenous leukemia	<i>Alpha streptococci</i> , <i>Neisseria catarrhalis</i> <i>Proteus mirabilis</i>
2	Limongelli et al. ⁽⁶⁾	1976	Noma-like	63/M	Chronic lymphocytic leukemia	<i>Bacterioides melangenicus</i>
3	Bendl et al. ⁽²⁾	1983	Noma	54/M	Lymphoma	<i>P. aeruginosa</i> , <i>S. aureus</i> , <i>Enterococcus spp</i>
4	Bendl et al. ⁽²⁾	1983	Noma	60/F	Chronic myeloid leukemia with iron deficiency	<i>P. aeruginosa</i> , <i>S. aureus</i> , <i>C. albicans</i>
5	Brady-West et al. ⁽³⁾	1998	Noma	30/F	T cell acute lymphoblastic leukemia	<i>P. aeruginosa</i> , <i>S. aureus</i> , <i>K.pneumoniae</i>
6	Present case	2010	Noma-like	17/M	Acute promyelocytic leukemia	<i>Enterococcus spp</i> , <i>S. aureus</i> and <i>C. albicans</i>

M = male; F = female

oris.⁽³⁾ This patient's lesion was diagnosed as a noma-like lesion because of the similarity of its clinical features: rapid progression to gangrenous necrosis of the hard and soft palate, uvula and right tonsil, leading to severe mutilation, as seen in noma lesions but of a different etiopathogenesis (*Enterococcus spp*, *Staphylococcus aureus* and *Candida SP* infection).⁽⁷⁾

The differential diagnoses are: mucocutaneous leishmaniasis; lupus erythematosus; leprosy; agranulocytic ulcerations; physical trauma (including burns); syphilis; oral cancer and yaws.⁽¹⁾ Infections of the oral cavity can spread to other parts of the body.⁽¹⁾ The prognosis for nomas is largely dependent upon early and accurate diagnosis and treatment.

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