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LETTER TO THE EDITOR

An unusual cause of facial wound in a child: Hyper IgE syndrome-associated Noma neonatorum

A 4-month-old failure to thrive (FTT) infant presented with large ulcerative ecthyma-like lesions on the face. Five days before the well-developed facial wound, a small area of erythematous papule and plaque was appeared on the right cheek and around the nose. She was febrile, restless, and breastfeeding intolerant. Fever was started 2 days before cutaneous lesions showed up. Lesions started at the cheek with facial oedema and progressed in 2 days to a large ulcerative area on the upper lip, right cheek, and around the nose (Figure 1). The child had a history of similar lesions when she was 2-month-old. She also had cervical and posterior scalp lymphadenopathy. Extensive eczematous lesions were seen on the trunk and limbs. On clinical and laboratory evaluation high serum levels of IgE and peripheral eosinophilia were detected. The clinical diagnosis of hyper IgE syndrome (HIES)–associated Noma neonatorum was made and she was undergoing it. She was treated with parenteral antibiotics for 2 weeks in combination with local wound care. The necrotic crust was gradually removed and scar formation remained at the site of infection. (Figure 2).

Noma neonatorum is a rare opportunistic, necrotising ulcerative infection that mostly affects malnourished and immunocompromised children.[1]

Necrotising stomatitis and gingivitis can extend rapidly to the maxilla and mandible and other adjacent surfaces, which appear as a well-demarcated black necrotic area. Many children do not survive this stage due to sepsis formation. In children who passed from the critically acute phase, the wound healing process will start with granulation tissue and extensive fibrosis formation that is associated with wound contracture with devastating facial deformities which potentially can result in physical impairments including limitation of mastication, oral incontinence, speech problems, and aesthetic disfigurements.[2,3]

There are several risk factors for Noma. Most of these risk factors can be related to poverty (especially in developing countries) including economic malnutrition, poor oral hygiene, poor sanitation, low vaccination coverage, and systemic infections (eg, malaria, measles). Another important category as a risk factor of NOMA is related to all conditions associated with reducing the host's immunity

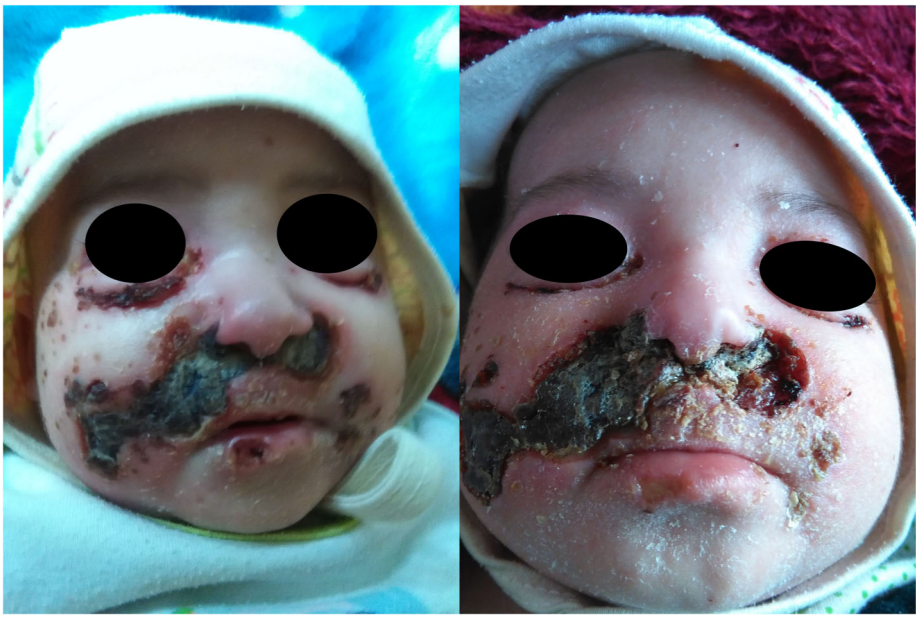

FIGURE 1 Noma neonatorum. Gangrenous ulcer involving mucocutaneous junctions of oral, nasal, and also cheek and eyelids in association with facial oedema

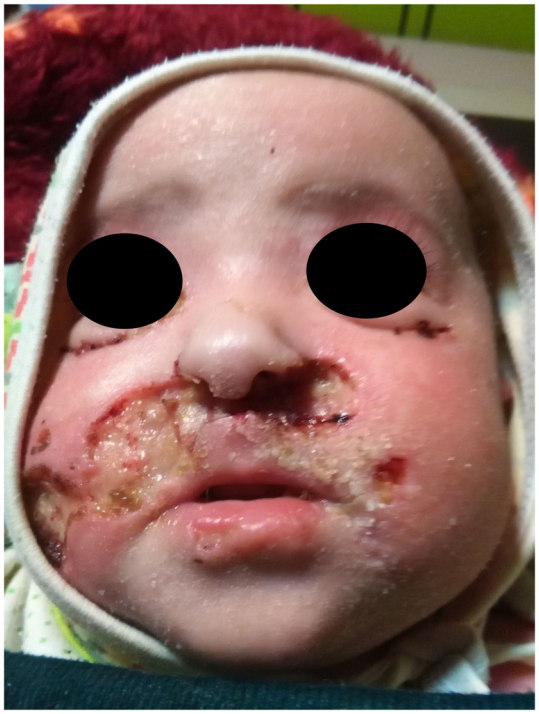

FIGURE 2 Noma neonatorum. Post-treatment scar formation

including primary and secondary immunodeficiency states.[4,5]

Over the past two decades, there has been an increase in the incidence of Noma in association with immunosuppressive therapy and patients with immunosuppressed conditions.[6]

NOMA has been reported in cases with human immunodeficiency virus (HIV)[7-9] and acquired immunodeficiency syndrome (AIDS),[10] severe combined immunodeficiency (SCID),[11] and chemotherapy-induced neutropenia.[12]

The current article is the first case of Noma in a neglected child with a recent diagnosis of HIES. In HIES, there is abnormal signalling in the pathway of Thelper-17, that results in immunodeficiency and susceptibility to mucocutaneous and sinopulmonary infections. Th17 has an important role in the recruitment of neutrophils for host defence.[13]

As Noma is fulminant in clinical course and within days can evolve from small ulcer to large necrotising wound if left untreated, clinical suspension and early diagnosis especially in the setting of immunodeficiency syndrome can prevent the major destructive and sometimes life-threatening sequels.

In addition to early identification and specific antimicrobial therapy, identifying risk factors and related conditions with proper treatment of the underlying diseases is important.[5,14]

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This research has been ethically approved and the Approval ID is: IR.ARI.MUI.REC.1400.114.

CONFLICT OF INTEREST

The authors have no conflicts of interest to declare.

DATA AVAILABILITY STATEMENT

the data that support the findings of this study are available on request from the corresponding author.

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