Periocular Necrotizing Fasciitis

Maarten P. Mourits

Learning Objectives

• The diagnosis of necrotizing fasciitis can be troublesome, but an early start of treatment is essential to prevent death. So, in case of doubt, go for the worstcase scenario.

Introduction

The infection, which sometimes starts at a small skin lesion (varicella for example) or after surgery, spreads along the fascia of the muscles and causes necrosis of the skin, subcutaneous fat, fascia, and sometimes the underlying muscles. In common parlance, bacteria causing NF are therefore called "flesh-eating bacteria." NF is a rare disease in which a multitude of factors play a role. Large series of patient cases are lacking and hence reports on incidence, etiology, prognosis, mortality rate etcetera are often conflicting. NF seems to be more common in men than in women. It occurs more frequently in African and Asian countries. It is extremely rare in children.

Many risk factors (diabetes mellitus, burns, intravenous drug use, varicella, immunosuppres-

M. P. Mourits (🖂)

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Amsterdam University Medical Centers, Location AMC, Amsterdam, The Netherlands

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sion, malnutrition, age >60 years, decreased kidney function, malignancy, and arterial vasculopathy) have been suggested, but diabetes mellitus appears to be the most important. From a systematic literature search including 1463 patients, it is evident that in almost 50% of cases diabetes mellitus is a comorbidity [1].

NF must be differentiated from erysipelas (which has a less fulminant course) and gas gangrene by means of culturing micro-organisms from infected tissues. In addition, orbital cellulitis must be distinguished from PONF. Signs that help to come to a right and early diagnosis are pain that is out of proportion, failure to respond to broad-spectrum antibiotics, and the presence of bullae and necrosis [2].

Presentation

Necrotizing fasciitis (NF) is a life-threatening bacterial or fungal infection of the subcutaneous tissues. Most often, the extremities and the perineal region are involved [1], but occasionally the eyelids and other parts of the face are the target of the infection (Figs. 21.1, 21.2, 21.3 and 21.4). In that case, we speak of periocular necrotizing fasciitis (PONF). Characteristic for the disorder is extreme pain together with swelling and redness of the skin. Within hours to days, the area turns from red to purple and then to black. Bullae may arise, and crepitations can be



21

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Fig. 21.1 and 21.2 A 68-year-old male with PONF of the left part of his face starting around the orbit at admission and 72 h later after debridement



Fig. 21.3 and 21.4 A 84-year-old male with PONF of the left orbit after debridement and after rehabilitative surgery

felt. The inflamed area is relatively painless, whereas the adjacent parts are extremely painful. Next, the patient becomes seriously ill and somnolent with high fever, nausea and diarrhea, leukocytosis, and high sedimentation rates. Release of toxins can cause toxic shock syndrome associated with multi-organ failure an acute respiratory distress syndrome. Without treatment, the disease is fatal within 24–72 h.

Etiology

Four types of NF can be distinguished as follows

 Type 1: Caused by a mixed flora of anaerobic and aerobic bacteria. This type is seen most often in patients with diabetes mellitus. Crepitations may be present. It is assumed that 70–80% of all cases belong to this type. The prognosis is relatively good.

- 2. Type 2: Caused by group A β-hemolytic Streptococcus (i.e., *Streptococcus pyogenes*) and by many other types of bacteria. *Streptococcus pyogenes* is found in 20–30% of cases of NF, but in up to 85% of patients with PONF [3]. This type of NF is especially seen in previously healthy individuals and is known for its fulminant course with toxic shock syndrome. Untreated, it is almost always lethal. However, PONF caused by *Streptococcus pyogenes* has a much better prognosis.
- 3. Type 3: Caused by Vibrio species from seawater. This type is seen especially in Asian countries and has a high mortality rate.
- 4. Type 4: Caused by fungi (e.g., Candida). This type also has a high mortality rate.

Treatment

Treatment initially consists of intravenous administration of broad-spectrum antibiotics and surgical removal (debridement) of the necrotic tissues. Every 2 h the boundaries of the inflamed tissues have to be demarcated with a dermo-marker in order to follow progression of the disease. Because of obliteration of blood vessels and tissue necrosis, the antibiotics not always reach their targets and this explains the initial failure of medical treatment. So, it is important to remove the necrotic tissues until healthy tissue is attained. This must be repeated until the inflammation is gone. It is not exceptional that at least five sessions are required [3]. It is evident that antibiotic treatment should be adjusted accordingly as soon as the causative pathogens have been identified. In addition to local measures, general (hemodynamic, respiration) measures must be taken [4]. Once the inflammation is gone, reconstruction of the tissue defects with free or pedicled flaps can be undertaken (Fig. 21.4).

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