Necrotising fasciitis

Necrotising fasciitis is a severe, rapidly progressing, and invasive soft tissue infection caused by bacteria, most commonly Streptococcus pyogenes, Group A streptococci (GAS).

Necrotising fasciitis (necrosis = tissue death, fasciitis = inflammation of the tissues surrounding muscles, nerves, fat, and blood vessels) is a rare invasive bacterial infection that affects the subcutaneous tissue and the muscle sheath. It destroys deep soft tissues, including muscle sheath and subcutaneous fat, and rapidly spreads throughout the body, potentially leading to fatal outcomes without prompt and proper treatment.

Necrotising fasciitis can affect any part of the body, but it usually occurs on the extremities, and in two-thirds of patients on the legs. In children, it most commonly affects the abdominal wall, extremities or perineum.

The cause

Necrotising fasciitis is a polymicrobial necrotising soft tissue infection caused by various bacteria. The most common causative agent is Streptococcus pyogenes (beta-haemolytic group A streptococcus), but it can also be caused by Gram-negative bacilli enterococci, staphylococci, and anaerobes.

Reservoir

The reservoirs for Gram-negative bacilli, enterococci, staphylococci, and anaerobes are humans, animals, and the environment. However, for Group A streptococci (GAS), humans are the only reservoir. In humans, S. pyogenes is commonly found in the nasopharynx and on the skin.

Transmission

In necrotising fasciitis, the initial entry of bacteria into the body can occur through several routes. In the case of open skin lesions or wounds, bacteria from the environment can pass directly to the site of infection. Skin injuries may include: cuts, abrasions, burns, insect bites, puncture wounds (including those caused by intravenous drug use) and surgical wounds.

Infection can also occur with blunt trauma that does not damage the skin, e.g. bruises and muscle strains. In blunt skin injuries, the most likely mechanism of infection is transient bacteraemia (the presence of bacteria in the blood).

The inflammation then typically spreads from the subcutaneous tissue deep into the muscle sheaths.

Risk factors

Anyone can get streptococcal necrotising fasciitis. It is extremely rare, most commonly found in people with systemic or local immune deficiency.

Risk factors for necrotising fasciitis include diabetes mellitus, peripheral vascular disease (thickening and subsequent narrowing of the blood vessels due to fat accumulation), treatment with immunosuppressive drugs, obesity and cirrhosis of the liver.

In children, those at higher risk of developing the disease include new-borns with omphalitis (inflammation of the umbilical cord) and balanitis (inflammation of the skin on the penis), which are most often complications of circumcision, children with congenital immune system disorders, children with Down syndrome, those with cancerous diseases (leukaemia, rhabdomyosarcoma), poorly controlled diabetes mellitus, and children on immunosuppressive therapy, especially with glucocorticoids. The disease can also affect healthy children and may occur as a complication of chickenpox, skin injuries, surgical procedures, or blunt soft tissue trauma.

Incubation

The incubation period (the time between infection and the first symptoms/signs of disease) is short, lasting a few hours, rarely several days. Usually, the disease develops within the first 24 hours after the injury.

Clinical picture

Disease symptoms and signs characteristic of necrotising fasciitis are the following:

- Early symptoms and signs develop quickly (within the first 24 hours):
  - Red, warm, swollen and tender skin lesions that spread rapidly;
  - Intense pain, disproportionately severe in relation to signs of local infection.
  - Chills and fever.

- Advanced symptoms and signs (usually occurring within 3–4 days) most often include:
  - Ulcers, blisters or black patches on the skin;
  - A dark red sore;
  - Changes in skin colour (skin may turn black within 24–48 hours);
  - Oozing or discharge from the infected area;
  - Tiredness and dizziness;
  - Nausea and diarrhoea.

In necrotising fasciitis, the clinical course of the disease is extremely rapid, and irreversible septic shock with multi-organ failure and death can occur within a few hours after injury.

Primary skin lesions (minor abrasions, insect bites, injection site, impression) are present in about 80% of patients' medical history (information given by the patient about their problems before starting treatment). The infection usually spreads from the site of the primary skin lesion or the operated area.

In children who have chickenpox, infection of the existing rash caused by varicella zoster virus with bacteria that cause necrotising fasciitis can occur. In these children, cutaneous signs of necrotising fasciitis may not be present. However, a high fever may persist for 3–4 days after the onset of chickenpox symptoms, or a fever that has already subsided may return.

Complications and outcome of the disease

Serious complications are common in necrotising fasciitis. Complications occur due to the spread of bacteria to nearby and/or distant tissues and organs very early in the course of the disease. Necrotising fasciitis can be complicated by:

- Organ failure;
- Streptococcal toxic shock syndrome (serious infection with pyogenes, which secrete exotoxins that damage tissues, enter the blood and can lead to sepsis, multi-organ failure and death);
- Sepsis (an extreme systemic response of the body to an infection that triggers a chain reaction of tissue damage, organ failure and can lead to death).

It can also cause lifelong complications due to severe scarring or limb loss due to surgical removal of infected tissue or amputations.

Despite prompt recognition and appropriate treatment, necrotising fasciitis is associated with a high mortality rate. The mortality rate is 20–47% in adults and up to 60% with associated complications. The mortality of affected children with associated complications is 70%.

Diagnosis

Diagnosis must be made as early as possible, as any delay in initiating treatment greatly increases mortality.

The diagnosis is made based on a characteristic clinical picture, laboratory tests, microbiological investigations and surgical examination of the affected area.

Sometimes there are no changes on the skin because the infection is spreading in the subcutaneous tissue, making it difficult to diagnose the disease.

Laboratory tests reveal an increased number of leucocytes with a high proportion of immature neutrophilic leucocytes. Additionally, there is usually a decrease in the number of lymphocytes in the blood. C-reactive protein is elevated. It is possible to observe decreased platelet counts, anaemia, reduced concentrations of calcium and sodium in serum, elevated plasma bilirubin, as well as increased aminotransferases and creatinine.

The tissue sample must be sent for microbiological examination (Gram stain, bacteriological culture) and histological examination (frozen section). The cause is confirmed by isolating bacteria from the inflamed tissue or from the blood.

Imaging studies such as ultrasound (US), computed tomography (CT), and magnetic resonance imaging (MRI), which provide deep views of the body or organs in layers, can show soft tissue swelling, thickening of the muscle fascia, and potentially the presence of air in the tissue.

Differential diagnosis

Some other soft tissue infections have a similar clinical picture, such as:

- Anaerobic streptococcal myositis (muscle inflammation; slower progression and less swelling, but some redness);
- Gangrene in peripheral vascular disease (tissue death from thickening and subsequent narrowing of the vessels due to fat accumulation; slow progression of the disease);
- Concomitant necrotising cellulitis (inflammation of the subcutaneous tissue with local tissue death; occurs 3–14 days after infection).

There are many infections that, especially in the early stages, resemble necrotizing fasciitis, which can complicate diagnosis. Rapid intervention is crucial, so treatment should be initiated as soon as possible, despite any incomplete investigations.

Treatment

Necrotising fasciitis is a very serious condition that requires hospital care. It is treated with a combination of antibiotics and surgery. It is essential to remove the dead tissue immediately and to establish wide drainage, which must be repeated several times a day if necessary.

Initial antibiotic treatment is experimental and targeted against the most common pathogens. The duration of antibiotic treatment is not precisely defined and is required as long as local and systemic signs of infection are present or repeated surgical procedures are necessary.

Chemoprophylaxis

Chemoprophylaxis aims to protect people from infection and disease with the right medicines. Close contacts of a patient with an invasive GAS infection can be ordered appropriate antibiotic protection. Chemoprophylaxis is usually decided on an individual basis and only in people who are at increased risk of death if they develop an invasive infection (elderly, individuals with systemically or locally compromised immune systems such as those with diabetes, kidney disease, cirrhosis of the liver, cancer or peripheral vascular disease).

Prevention

There is no vaccine against necrotising fasciitis.

Maintaining good personal and general hygiene is important to prevent infection. In particular, good hand hygiene is important, especially when caring for wounds.

To prevent necrotising fasciitis, it is important to:

- Clean all minor cuts and injuries that wound the skin (such as blisters and abrasions) with soap and water;
- Clean and cover open wounds with clean, dry bandages until they have healed;
- Deep or extensive wounds require medical attention;
- Treat fungal infections such as athlete's foot;

- In case of open wounds or skin infections, avoid spending time in hot tubs, swimming pools and natural waters (lakes, rivers, sea).

Infectiousness

Necrotising fasciitis is rarely infectious. Most cases of necrotising fasciitis occur accidentally after an injury and it is very rare for the infection to be passed on to other people.

Immunity

Immunity does not develop. Rarely, a person who has had necrotising fasciitis may become ill again in the future.