Case report

FOURNIER'S GANGRENE AS MANIFESTATION OF ANOGENITAL NECROTIZING FASCIITIS

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SUMMARY

The authors present the main clinical features, pathogenesis and therapy in a typical case of Fournier's gangrene in a genital localization. This foudroyant infection with high mortality is regarded as a characteristic form of a toxic shock-like syndrome manifested as a necrotizing fasciitis. It is emphasized that despite adequate therapy certain basic diseases or predisposing factors can still result in a lethal outcome.

Keywords

Fournier's gangrene, necrotizing fasciitis, toxic shock-like syndrome

INTRODUCTION

At present, Fournier's gangrene is generally defined as a hyper-acute necrotizing infection with high mortality, which involves subcutis and fasciae of the anogenital area. The first case of infection which affected male genitals was described in 1883 by the French dermatologist Fournier. Later, perineal, ano-rectal and vulval forms were also reported. In the past, different terms have been used to describe the disease: synergic gangrene non-clostridial myonecrosis of the perineal and scrotal skin; perineal phlegmone.

A typical case history of Fournier's gangrene with lethal out-come, which developed as a sequel to multiple underlying diseases is presented along with its clinical features, pathogenesis and therapy.

CASE HISTORY

A male patient aged 55 had a history of malignant hypertension, cardiac decompensation, chronic glomerulonephritis, and spleno-megaly (myeloid metaplasia). In 1995 the patient was admitted to a department of medicine for progression of hepato-splenomegaly, aggravation of myocardial lesions due to hypertension, generalized edema due to deterioration of renal function, and erysipelas affecting the lower right limb. Following antibiotic therapy (penicillin) the erysipelas healed, but some days later a painful, edematous erythema with circumscribed skin necrosis and subsequent suppuration of the scrotum developed. At that time the patient was admitted to our Department.

Genital status at admission: light-red discoloration of

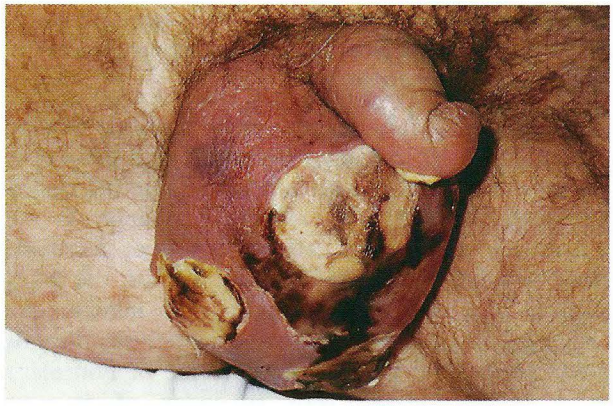

Figure 1. Edema and incipient necrosis of scrotum.

the scrotum and penis with livid-red hemorrhages and edema. Yellowish-brownish necrotic areas with clear demarcation could be seen on the scrotum and the external surface of preputium; bilateral macerated scaly erythema with superficial fissures was observed in the inguinal region (Fig. 1).

General status: generalized edema, emphysematous thorax, signs of diffuse congestion of the basal part lung, enlarged heart (3 cm to the left); aortic ejection murmur of a 2/6 intensity, protruding abdomen, ascites, hepatomegaly reaching the level of the navel, and compact splenomegaly reaching to the line of the crista iliaca.

Laboratory tests: RBC sedimentation 58 mm/h; hematocrit 0,28; hemoglobin 8,2 mmol/l; WBC 15100 G/l. Urine: no pathological findings. Total serum (se) protein 55 g/l; res. nitrogen 17,4 mmol /l; se creatinin 260 µmol/l; blood glucose 4,1 mmol/l; se bilirubin 22 µmol/l; SGOT 11 U/1; SGPT 7 U/l; gamma-GT 550 u/l (up to 50); ALP 139 u/l (up to 280); LDH 683 U/l (up to 460).

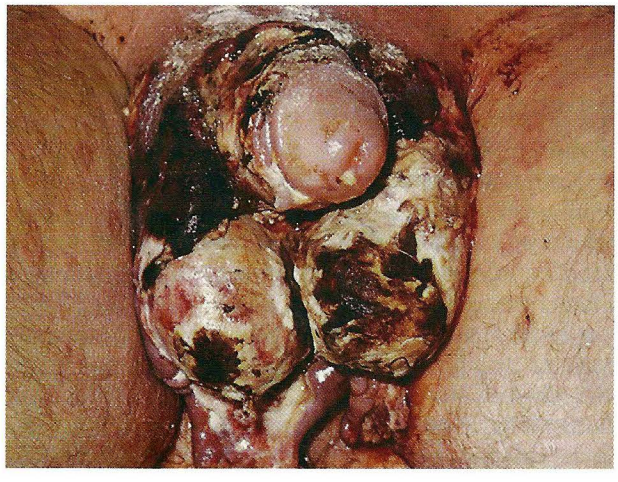

Figure 2. Extensive necrosis of scrotum and penis.

Ultrasonic examination of the genitals: sustained fluid accumulation in the scrotum around both testicles. Homogenous left testicle. An inhomogenous, echo-poor cystic area of 2 cm in diameter in the right testicle. Adequate blood flow bilaterally.

Bacteriology of the necrotic scrotal tissue: Enterococcus, Proteus, Pseudomonas aeruginosa, Streptococcus pyogenes.

Course of the disease and therapy:

The diagnosis of Fournier's gangrene was based on the clinical findings. The necrotic scrotal and preputial tissue was resected under narcosis. The necrotic fascia of the entire scrotum was also removed. Visual analysis and palpation of the testicles suggested intact tissue. The perineal surgical wound was left open and was drained.

Histological findings: subepidermal granulation tissue rich with vessels and cells; extensive loci of hemorrhagic necrosis containing unstructured pus cells.

Considering the heterogeneity of bacterial infection, a high-dose complex antibiotic therapy (Ampicillin and Netromycin, then Fortum and Netromycin, later Rocephin and Ciprobay) was started. The therapy was supplemented with metronidazol (Klion) infusion and local Betadine dressings. Despite the above-mentioned treatment and supplementary therapy, deterioration of the general status and aggravation of the multiple internal organ insufficiencies (respiratory, hepatic, and renal symptoms) developed. New necrotic areas appeared on the scrotum. The patient rejected another surgical intervention and asked to be transferred to the hospital where he had first been admitted. Several days later the patient died in septic shock.

DISCUSSION

Fournier's gangrene typically occurs in male patients around the age of 50, although the disease has been reported in all age groups including children (1). In most of the patients certain predisposing factors can be found: 1. urologic interventions (e.g. prostate puncture) or diseases (periurethral sepsis); 2. diseases of the perineal area and the rectum (abscesses, fistulas); 3. pathological processes affecting the internal organs (alcohol abuse, obesity, diabetes

mellitus, circulatory disorders, tumors, and recently HIV infection). The entry point for the infection may be identified in all cases, but being minor or insignificant, it usually remains undiscovered (mechanical devices used for treatment of impotence are reported as a new source of infection) (2,3,4).

As a rule, the infection is caused by multiple pathogens. In 70% of the cases the infection is of mixed character, in 20 % it is solely aerobic, and in 10 % anaerobic. The most typical pathogens are E. coli, Streptococcus pyogenes, Pseudomonas aeruginosa, Klebsiella pneumoniae, Proteus mirabilis, and anaerobic Streptococci. More rarely, in young patients the infection was identified with the A-group Streptococci (2).

The prodromal symptoms are extreme pain of the affected skin area and edematous erythema; the severe pain shows no correlation with the observed clinical features. The cutaneous symptoms represent only the "tip of the iceberg", since the infection has penetrated along the fasciae into the deep tissues. 48 hours following the appearance of erythema, the affected area becomes bullous and necrotic; later suppuration develops, which is accompanied by general symptoms (fever, prostration). Crepitation is often present. In the absence of adequate therapy the pathologic process spreads into the abdominal wall and the gluteal region (5).

At present, Fournier's gangrene is considered to be an anogenital form of necrotizing fasciitis (type I) (2,6). The infection is caused by heterogeneous flora (obligatory and facultative anaerobic microorganisms). Regional intravascular coagulation due to bacterial toxins and insufficiency of the protein C and S systems significantly contributes to the development of severe local symptoms (5,7) and tissue hypoxia; the latter leads to further spreading of the gangrene. There is evidence that the local Schwartzman effect may play a role in the development of the gangrene (8). At present, necrotizing fasciitis and Fournier's gangrene are regarded as a toxic shock-like syndrome with characteristic hypotension, fever, destructive infection of the soft tissues, and multisystemic internal organs insufficiency (liver, kidney, respiratory system); hemoculture is always positive (9). It is generally agreed that the toxins and the enzymes of invasive pathogens cause this severe clinical picture (6,9).

The diagnosis of Fournier's gangrene is based on clinical features, modern imaging methods (MRI), and studies of the frozen sections made prior to surgical intervention. The intraoperative picture verifies the diagnosis. The infectious gangrene should be differentiated from erysipelas, cellulitis, ischemic vascular necrosis, gangrenous ecthyma, ulcerous pyoderma, and coumarol necrosis (10).

The mortality rate of the disease is high, ranging from 20 to 50%. An age of over 50, and laboratory findings indicating insufficiency of the internal organs aggravate the prognosis. Although opinion is not unanimous on this point, survival is further decreased when the affected area exceeds 5% of the body surface. It is also noteworthy that the number and timing of surgical interventions does not significantly affect survival (at present the outcome of Fournier's gangrene is assessed with an index of severity; values under 9 indicate better prognosis) (6).

Similarly to necrotizing fasciitis, there are three basic principles in the therapy of Fournier's gangrene: antibiotic treatment, immediate surgical intervention and necrectomia, as well as supplementary therapy. In regard to the mixed character of the infection, therapy should be initiated with amoxycillin + clavulinic acid combined with high dose metronidazol. Alternatively, therapy with aminoglycosides or clindamycin may be considered. Necrectomia should be performed until the entire devitalized tissue has been removed (bleeding tissue appears). Castration is unnecessary. Patients should be treated in an intensive care unit, where adequate electrolyte and fluid replacement, as well as cardiorespiratory support are provided. The recently introduced systemic steroid therapy (local Schwartzman effect), intravenous administration of gamma-globulin, as well as pentoxiphyllin treatment which inhibits accumulation and action of endogenous cytokines (T-cell activity induced by super-antigen) can be considered as adjuvant therapy. The defect can be reconstructed with plastic surgery (free transplantation, fasciocutaneous graft (11,8)).

CONCLUSION

The presented case history is a typical example of Fournier's gangrene. Perineal intertrigo and mycosis served as an entry portal for the infection. Predisposing factors, such as cardiac insufficiency, chronic glomerulonephritis, and myeloid metaplasia further aggravated the course of the infection, and finally lead to multiple internal organ insufficiency. Albeit the surgical intervention was performed several days later, data from the literature indicate that the lethal outcome resulted from the underlying diseases and the severity of the infection. We failed to verify a better prognosis when less than 5 % of the body

surface was affected, because the involvement of deep and remote tissues can be much more extensive than the gangrene visible on the surface. The presented case demonstrates the extreme severity and often lethal outcome of Fournier's gangrene. This case emphasizes the importance of differential diagnosis of the inflammatory cutaneous symptoms in the anogenital region, where the possibility of Fournier's gangrene should always be considered. Early recognition and comprehensive therapy of the disease is of utmost importance.

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