

Case Report

Fournier gangrene associated with hyper IgE syndrome (Job syndrome)

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Abstract: We report a case of a 32-year-old man with hyper IgE syndrome (Job syndrome) who developed Fournier gangrene due to infectious multiple atheromas of the scrotal skin that progressed to the right groin and thigh. The patient required surgical debridement and subsequent skin grafting. This is a rare case of Fournier gangrene associated with hyper IgE syndrome (Job syndrome). When a patient without diabetes mellitus has repeated infections and atopic-like dermatitis, Job syndrome should be considered.

Key words: Fournier gangrene, job syndrome, surgical debridement.

Introduction

Fournier's gangrene is a necrotizing fasciitis of the male genital region that can become critical.¹ In 1883, Fournier first described it affecting the perineal and genital regions.^{1,2} On one hand, Hyper IgE syndrome is defined as a primary immunodeficiency disease that presents with recurrent *Staphylococcus aureus* infections, refractory eczema from infancy, and marked elevation of serum IgE levels.³ In 1966, Davis *et al.* called this syndrome 'Job syndrome' based on the notion that Job of the Old Testament had multiple cold abscesses.³

Herein, we report a case of Fournier's gangrene associated with Job syndrome.

Case report

A 32-year-old man was admitted due to multiple scrotal infectious atheromas. He visited the outpatient dermatology clinic and was given oral antibiotics. One week after admission, the patient returned to the dermatology clinic complaining of scrotal and femoral pain. At that time, the patient had erythema and a high fever, and he was admitted to the urology department with a diagnosis of Fournier's gangrene (Fig. 1). Laboratory data revealed an increased white blood cell count (22 920/mm³), an increased C-reactive protein (19.57 mg/dL) and an increased serum IgE (10 000 IU/mL). Positive reactions for *Staphylococcus aureus*- and *Candida albicans*-specific IgE antibodies were also noted. His past history included many different infections and atopic-like dermatitis. For example, at the age of 1 year, he developed septicemia secondary to osteomyelitis of the lower left thigh. In addition, since childhood, he had atopic dermatitis. At 31 years of age, the patient required incision and drainage of a cervical lymphatic abscess. He had no history of diabetes mellitus. He had laboratory data typical of the syndrome, such as an elevated IgE; furthermore, the patient had a characteristic face, with a prominent forehead and a wide nasal septum (Fig. 2). The patient was diagnosed as having Fournier's gangrene associated with hyper IgE syndrome (Job syndrome). He required surgical debridement of the scrotum, right groin, and right

thigh to remove devitalized tissue (Fig. 3). The spermatic cords and testes were not affected by the infection. *Methicillin-resistant Staphylococcus aureus* (MRSA) and *Peptostreptococcus micros* were isolated. The wound was intentionally not closed, and the open wound was irrigated with saline daily. After the infection subsided, fresh granulation tissue grew. Finally, a skin graft from the anterior femoral region was carried out, and the wound healed completely.

Discussion

Generally, the overall mortality rate of Fournier's gangrene ranges from 15% to 50%, and this is an acute urological emergency.² The underlying diseases include diabetes mellitus, alcoholism, internal malignancy, severe infection, and immune system deficiency. Mixed infection with aerobes and anaerobes is mainly present. The most common isolated bacteria are *Escherichia coli*, followed by *Bacteroides* and *Streptococcal* species.² The lesions tend to be confined to the subcutaneous tissue, the skin, and Colles' fascia; there are a few cases in which the inflammation spreads to the spermatic cords and the testes.² Urgent surgical debridement of the inflammatory tissue is the most important treatment. Simultaneously, the adequate use of antibiotics and control of other complicating factors, such as diabetes mellitus, are also required.^{1,2} When natural closure of the wound cannot be expected, it is kept open. Subsequently, the wound is closed in two stages.^{1,2}

The case reported here is a rare case of Fournier's gangrene associated with hyper IgE syndrome (Job syndrome), which is a primary immunodeficiency disease. The main clinical findings in Job syndrome patients include skin infections caused by *Staphylococcus aureus*, pneumonia, repeated lymphadenitis, mucocutaneous candidiasis, and marked serum IgE elevations.⁴ The genetics of the disease are thought to include an autosomal dominant inheritance in some instances.^{4–7} In Japan, 37 patients with Job syndrome have been reported.⁴ The present case of gas gangrene associated with hyper IgE syndrome is the second such reported case in Japan.⁴ It is clear that the main cause of this disease is a deficiency of interferon- γ production by helper T cells.⁵ In particular, the reason that patients with hyper IgE syndrome are commonly infected with *Staphylococcus aureus* and *Candida albicans* may be that IgE antibodies against these organisms induce type-I allergy, which inhibits neutrophil phylaxis.^{6,7} The clinical characteristics of this syndrome include atopic-like dermatitis, chronic cutaneous abscess, pneumonia, and

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Fig. 1 Macroscopic finding of the scrotal wound. Multiple infectious atheromas of the scrotal skin are necrotized.



Fig. 3 Surgical debridement of the scrotum and thigh was carried out.



Fig. 2 The facial characteristics of the patient.

otitis media. In addition, Job syndrome patients have particular facial features, referred to as 'coarse face'; these features include a thickened, doughy texture of the facial skin, a broad-based nose, and a prominent forehead.⁴ Job syndrome is difficult to diagnose, since

patients present with what appears to be a simple atopic dermatitis.⁵ The prognosis of this disorder depends on whether *Staphylococcus aureus* pneumonia can be prevented and there is no underlying internal malignancy.⁴⁻⁹ It has been reported that malignant lymphoma may occur in patients with Job syndrome and has a very poor prognosis.⁹ In the present case, the patient's symptoms could be treated successfully. However, the patient will need treatment to prevent recurrent infections and periodic examinations for malignancy.

We report a rare case of Fournier's gangrene associated with Job syndrome. When a patient without diabetes mellitus has repeated infections and atopic-like dermatitis, Job syndrome should be considered.

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