PYODERMA GANGRENOSUM IN BURNED PATIENT - CASE REPORT

Pioderma gangrenozum kod pacijenta sa opekotinama - prikaz slučaja

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Summary

Introduction. Pyoderma gangrenosum is a rare, chronic, destructive, ulcerating skin disease of uncertain etiology. It develops most frequently in patients between 25-45 years of age and affects both sexes equally. Case report. We present a case of pyoderma gangrenosum in a young female patient who sustained a burn injury of 40% total body surface area. She underwent four operations. She developed a wound infection and urinary infection during her hospital stay. By the end of hospitalization, the papules followed with coalesce of ulcerations formed on the previously epithelized areas of her legs. The patient complained of the intensive pain localized on these surfaces. Since pyoderma gangrenosum was suspected, a dermatologist was included in treatment. Therapy was initiated (methyprednisolone 60 mg per day intravenously) with gradual reduction of the dosage. The patient was discharged from hospital two weeks later with almost fully complete cicatrization and epithelization. Conclusion. Pyoderma gangrenosum is still difficult to be diagnosed in the absence of specific and sensitive diagnostic methods; however, it is crucial to be suspected as early as possible and to start treatment immediately. Multidisciplinary approach is essential for optimal results.

Key words: Pyoderma Gangrenosum; Burns; Wounds and Injuries; Skin Transplantation; Pain; Signs and Symptoms; Therapeutics; Early Diagnosis

Introduction

Pyoderma gangrenosum was first described and named by Brunsting, Goeckman and O’Leary in 1930. They believed that streptococcal infection was responsible for secondary cutaneous gangrene and hence the name pyoderma gangrenosum was given. Today it is known that it is a misnomer [1].

It is a rare and serious disease. It develops most frequently in patients between 25 and 45 years of age and affects both sexes equally. Diagnosis is set empirically by the method of exclusion.

Case Report

A 24-year old female was admitted with a severe burn injury of 40% total body surface area (TBSA).

The burn was a combination of full thickness and partial thickness depth of the skin. She sustained it during a seizure in the bath-tub with hot water. On admittance, the primary surgical revision of burns was done and appropriate therapy initiated. During the 92-day hospital stay, she had three more operations. She developed a wound infection and urinary infection during her hospital stay. By the end of hospitalization, the papules followed with coalesce of ulcerations formed on the previously epithelized areas of her legs. The patient complained of the intensive pain localized on these surfaces. Since pyoderma gangrenosum was suspected, a dermatologist was included in treatment. Therapy was initiated (methyprednisolone 60 mg per day intravenously) with gradual reduction of the dosage. The patient was discharged from hospital two weeks later with almost fully complete cicatrization and epithelization. 

Sažetak


Ključne reči: Pioderma gangrenozum; Opekotine; Rane i povrede; Transplantacija kože; Bol; Znaci i simptomi; Terapija; Rana dijagnoza
nuclear antibodies on HEP-2 cells) and results came out negative. Graft acceptance was low due to a poor contact with the underlying tissue. Because of the excessive bleeding after the first operation, low thrombocite level (resistant to therapy) continuation of the necrectomy of the residual subdermal and deep dermal burns was postponed. The patient’s general condition was very bad, with periods of intermittent fever, high procalcitonine level, C reactive protein level and alkaline phosphatase values. Other blood analyses were within the normal range. The patient was treated with topical application of silver sulfadiazine until the granulation tissue was formed. The staged excocleation of granulation tissue and skin grafting was done (the first one a month later and the second one two months later). The wound swabs were performed twice a week and coagulase-negative staphylococci, Staphylococcus aureus and Acinetobacter spp. were isolated. Urine cultures and blood cultures were negative during entire hospitalization. At the beginning of the third month of hospital stay, the swabs were negative. At about that time, the papules turning into the ulcers started to appear on the areas of previously fully accepted and consolidated skin grafts and healed donor sites of legs. The ulcers kept on conflating, forming larger and larger surfaces of skin defects (circumscribed defects in total of 2% TBSA) (Figures 1 and 2). In consultation with a dermatologist, a new immunological testing was done (anti-nuclear antibodies, immunoglobulin (IgG, IgM, and IgA) and the results were within the normal limits. A skin biopsy was performed. Histopathological analysis revealed no specific changes (some signs of inflammatory reaction with neutrophils were found). All of the above led to clinical suspicion of pyoderma gangrenosum. Therefore, any surgical therapy was ruled out. Therapy with intravenous methylprednisolone was introduced, the dose being 60 mg per day during three days; afterwards it was reduced to 40 mg per day during the next 10 days. Oral administration of methylprednisolone was continued with gradual dose reduction. The ulcers were simultaneously treated with greasy gauze and compresses with ethacridine lactate. The local status was considerably better, cicatrization and epithelization from the edges was almost complete and the patient was discharged and advised about out-patient care.

**Discussion**

Pyoderma gangrenosum is a chronic, destructive, ulcerating skin disease. Its incidence is very low, being approximately one person per 100,000. It can be associated with other diseases such as inflammatory bowel diseases (15%), arthritis (37%) and hematological malignances. In 50% of cases, it appears as isolated skin conditions [2]. It may involve other organ systems, where it manifests as sterile neutrophilic infiltrates. Most common extra-cutaneous localizations are the lungs, heart, central nervous system, gastrointestinal system, eyes, liver, spleen, bones and lymph nodes [3].

Pyoderma gangrenosum is a disease of uncertain etiology, but there are a few hypotheses concerning its development such as genetic factors (it has been suggested to be autosomal recessive disorder), immunological factors (where abnormalities in both humoral and cell-mediated immunity are responsible for its occurrence), and vascular factors (which suggest that pyoderma gangrenosum may represent a type of vascular disorder) [2].

Pyoderma gangrenosum is classified into four varieties: ulcerative (classic form), pustular type (in which pustules do not evolve into ulcers), bullous (mostly in patients with myeloproliferative disease)
and vegetative type (lesions are chronic and limited, non aggressive variant) [1].

Classic pyoderma gangrenosum, most frequently occurring as a variant, is typically localized on the legs (in 75% of cases) [2]. It begins as a small papule or a collection of papules, and when they break down, an ulcer is formed. The ulcers coalesce with necrosis in the central area. Pyoderma gangrenosum presents as a deep ulcer with the defined border violet or blue in color. The edges are often worn and damaged and the surrounding skin is erythematous and indurated [4]. Its appearance can be accompanied by pain and deterioration of general condition with fever, malaise, arthralgia and myalgia. The characteristic feature of pyoderma gangrenosum is a pathergy reaction but it is present in about 25% of cases [2].

Histopathology of pyoderma gangrenosum depends on the timing and site of biopsy [4]. Massive neutrophilic infiltration in the absence of vasculitis and granuloma formation may be considered suggestive of pyoderma gangrenosum [1].

Diagnosis of pyoderma gangrenosum is based upon the morphology of lesions, clinical course and the tentative presence of underlying medical condition associated with its higher occurrence. There are no pathognomonic laboratory tests, diagnostic methods or histopathological finding [5]. Pyoderma gangrenosum is often a diagnosis of exclusion, and thus presents many clinical challenges; therefore it is frequently misdiagnosed [6].

There are numerous skin conditions which can mimic pyoderma gangrenosum such as infections (ecchyma, herpes virus ulcers, deep mycoses, etc.), necrotizing systemic vasculitis (Wegener's granulomatosis, polyarteritis nodosa, rheumatoid arthritis, etc.), proliferative processes, reaction to drugs (warfarin, iodine, etc.), autoimmune diseases and exogenous tissue injury [5].

Treatment of pyoderma gangrenosum requires multiple modalities in order to reduce inflammation and create optimal conditions for wound healing and pain control [6, 7]. There is no gold standard in treatment of pyoderma gangrenosum [8]. It is essential to exclude other infectious disease before it starts because corticosteroids and immunosuppressant drugs are therapy of choice [1]. Most treatments are empirical and based on small series or local experience. Immunosuppression is the main goal in treatment and it is usually achieved with corticosteroids and cyclosporine. Prednisolone is a drug of choice and it is introduced with high dosage (60–120 mg). Cyclosporine is mainly used to reduce the dependence on corticosteroids or in situations when corticosteroids fail. Other immunosuppressant, such as azathioprine, tacrolimus and antitumor necrosis factor α agents, can also be used [3].

Topical therapy can sometimes be sufficient for early and mild manifestations. It comprises treatment with wet compresses, hydrophilic occlusive dressings, antimicrobial agents and topical corticosteroids [2]. Gentle debridement with Burrow’s solution, silver nitrate or potassium permanganate baths are important in local treatment. Aggressive surgical therapy and skin grafting should be avoided. It could be performed if the patient is on systemic corticosteroid therapy until both the donor and recipient area are healed [1].

The prognosis of pyoderma gangrenosum is generally good. It must be emphasized that it is prone to recurrence and as a consequence leaves a residual scaring. Death from pyoderma gangrenosum is rare but may occur due to underlying medical conditions or as a result of the therapy [3].

In this case, pyoderma gangrenosum was observed in a young female patient, a burn victim. The changes appeared on her legs, where the skin grafts were previously fully accepted and the donor sites healed. They were in form of papules evolving into ulcers which were merging, and the defect was getting bigger every day in spite of everyday topical treatment. The patient was complaining of pain in this region. Extensive burns lead to disturbance in the cell mediated as well as in humoral immunity which can be an explanation for the development of pyoderma gangrenosum in our patient. Growing defects in the form of ulcers preceded by papules, resistant to any form of topical therapy, on typical localization, which was also a site of surgical intervention, should arouse a suspicion of pyoderma gangrenosum. It was empirically confirmed that the patient suffered of pyoderma gangrenosum when the positive response was obtained with parenteral and afterwards per oral therapy with corticosteroids. Laboratory and histopathological analyses showed no signs which could help to make the diagnosis of pyoderma gangrenosum. It was made by excluding other conditions which might lead to similar clinical presentation.

**Conclusion**

Each growing ulcer localized on the site of surgical intervention, which does not heal or react to topical treatment, should arouse suspicion of pyoderma gangrenosum, particularly if the patient is complaining of pain and worsening of general condition and has some of medical conditions associated with pyoderma gangrenosum.

**References**

