CASE REPORT / OLGU SUNUMU

Pyoderma gangrenosum in a patient with Crohn's disease: Case report and a review of the literature

Crohn hastalığında piyoderma gangrenosum: Olgu sunumu ve literatürün gözden geçirilmesi

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ABSTRACT

Pyoderma gangrenosum is a rare neutrophilic noninfectious dermatose. Etiopathogenesis remains unclear, but in half of cases, there is an associated underlying disease. Inflammatory bowel disease is the most common underlying disorder. Systemic immunosuppressive or immunomodulator drugs and some topical agents are used in treatment of pyoderma gangrenosum. Systemic corticosteroids are the first-choice of treatment. We reported a case with Crohn's disease associated with pyoderma gangrenosum. She was successfully treated with oral methyl prednisolon. The case was a 54-year-old woman who admitted to hospital because of erythematous, painful plaques on the right and left pretibial surfaces. She had a history of Crohn's disease, diabetes mellitus, and hypertension. An elevated white blood cell count (13500/μL) and high erythrocyte sedimentation rate (120 mm/h) were detected. A regime of broad-spectrum antibiotics was started, but response was poor. Histopathological assessment of biopsy specimens showed necrosis, severe edema and erythrocyte extravasations in superficial dermis, regenerative changes in adjacent epithelium, and mixed inflammatory reaction surrounding necrosis in the inner part of the dermis. Based on these clinical and laboratory findings, poor response to antibiotics and underlying disease; her skin lesions were considered as pyoderma gangrenosum. Oral methylprednisolone was started and her skin lesions improved. The steroid dose was tapered and finally stopped under outpatient follow-up. In conclusion, our patient also showed that corticosteroids continue to be the first-choice therapy in pyoderma gangrenosum.

Key words: Pyoderma gangrenosum, Crohn's disease, corticosteroids.

ÖZET

Piyoderma gangrenozum seyrek görülen ve enfeksiyöz olmayan bir nötrofilik cilt hastalığıdır. Etyolopatogenezi bilinmemektedir. Olguların yarısında altta yatan bir hastalık mevcuttur. En sık inflamatuvar bağırsak hastalığına eşlik eder. Tedavisinde immün baskılayıcı veya immünmodülatör ilaçlarla bazı topikal ajanlar kullanılmaktadır. Sistemik kortikosteroidler, tedavide ilk seçenek ilaçlardır. Crohn hastalığı zemininde gelişmiş ve başarılı bir şekilde tedavi edilmiş olan bir piyoderma gangrenozum vakası sunuldu. Crohn hastalığı, diyabetes mellitus ve hipertansiyonu olan 54 yaşında bir kadın, her iki bacağın ön iç tarafında kırmızımsı ve ağrılı lezyonları nedeniyle hastanemize başvurdu. Anormal laboratuar bulgusu olarak lökositoz (13500/μL) ve artmış sedimantasyon hızı (120 mm/saat) tespit edildi. Geniş spektrumlu antibiyotik tedavisi başlanıldı, ancak beklenen cevap alınamadı. Lezyon biyopsisinden histopatolojik değerlendirme yapıldı. Üst dermiste nekroz, şiddetli ödem, eritrosit ekstravazasyonuyla çevre dokuda rejeneratif değişiklikler, dermisin iç bölümünde nekrozu kuşatan mikst inflamatuvar reaksiyon görüldü. Kliniği, laboratuvar bulguları ve altta yatan hastalığı piyoderma gangrenozumu düşündürdü. Oral metil prednizolon başlanıldı, lezyonlarında düzelme görülmesi üzerine taburcu edildi. Ayaktan takiplerinde kortikosteroid dozu tedricen azaltılarak kesildi. Vakamız, sistemik kortikosteroidlerin piyoderma gangrenozum tedavisinde hâlâ favori ilaçlar olduğunu göstermiştir.

Anahtar kelimeler: Piyoderma gangrenozum, Crohn hastalığı, kortikosteroid.

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INTRODUCTION

Pyoderma gangrenosum (PG) is a severe ulcerative non-infectious neutrophilic skin disorder. The etiology is not clear. About 50% of patients have an underlying disorder. Inflammatory bowel disease is the most common underlying disease associated with PG. Other important associated systemic diseases include hematologic and rheumatologic conditions.[1,2] We report a demonstrative case of 54-year-old woman who have Crohn's disease accompanying pyoderma gangrenosum.

CASE REPORT

The patient was admitted with erythematous, painful plaques on the right and left pretibial surfaces. She had a history of Crohn disease, diabetes mellitus, and hypertension. On examination medial parts of the pretibial surfaces was swollen and tender with erythematous plaques. The lesion on right leg is larger than other lesion (Figure 1).

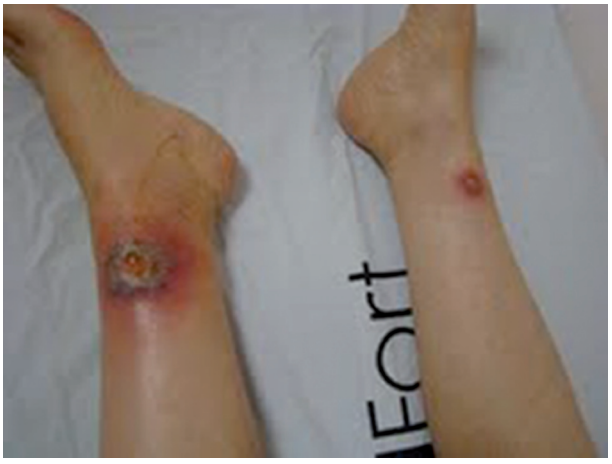

Figure 1. Appearance on the first days of admission to hospital

Laboratory investigations revealed an elevated white blood cell count of 13500/μL and a high erythrocyte sedimentation rate (120mm/h). The lesion was thought to be cellulitis, and a regime of broad-spectrum antibiotics was started. Multiple biopsy specimens were taken from the edge of the ulcer because of the poor response to the antibiotics.

Histopathological assessment revealed necrosis in superficial dermis, regenerative changes in adjacent epithelium, severe edema and erythrocyte extravasations in the superficial dermis and mixed inflammatory reaction surrounding necrosis in the inner part of the dermis. Bacterial cultures of the biopsy material and blood were negative for acid-fast bacilli, bacteria, and fungi.

The results of the following investigations were either normal or negative: blood biochemistry tests, liver function tests, hepatitis B and C, HIV, serologic tests, complement levels and antinuclear antibody test. Pyoderma gangrenosum associated with Crohn's disease was diagnosed. Methylprednisolone 64 mg/day was given initially for 5 days. A dramatic response occurred with a reduction of pain and erythema. Her lesions improved gradually, and she was discharged with a regime of prednisone 48 mg/day (Figure 2).

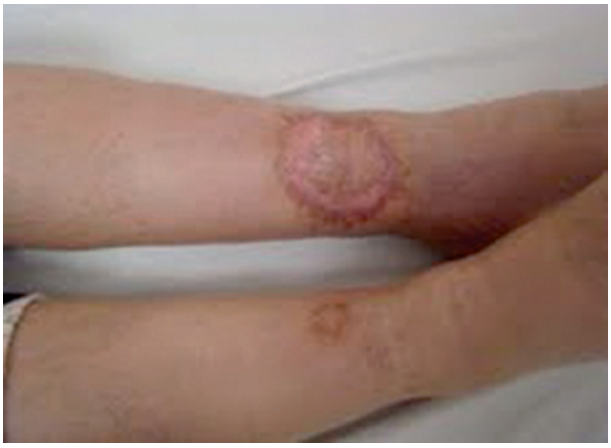

Figure 2. Appearance at discharge from hospital

DISCUSSION

Pyoderma gangrenosum is an uncommon necrotizing and ulcerative skin disease. The disease was first identified in 1930.[1] Since specific histopathological or immunofluorescent patterns are absent, the diagnosis is made clinically.[2] The earliest symptom may be localized pain, followed by small erythematous papules. The lesions rapidly evolve into tender pustules surrounded by indurated erythematous skin that breaks down to form an ulcer. The hallmark finding in pyoderma gangrenosum is rapidity of the development of the lesions and the painful ulcers with sharply circumscribed and demarcated, frequently undermined, livid borders and a necrotic base, and distinguish it from soft tissue infection.[3]

Lesions are most commonly found on the lower extremities but have been reported on the scalp, face, trunk, and arms.[2] Lesions may be single or multiple

and may be precipitated by trauma (pathergy). The Behçet's and the Sweet's syndromes also show this type of pathergy. Peak incidences occur in the third and fourth decades of life for female patients and in the fifth decade of life for male patients.[3] Pyoderma gangrenosum is a marker of various systemic diseases.[4] It is most often associated with inflammatory bowel disease. It occurs in 1% to 10% of patients with ulcerative colitis and in 0.5% to 20% of patients with Crohn disease.[5] The prevalence of PG in inflammatory bowel diseases was reported to be 2.3% inflammatory bowel disease in Turkish patients.[6] Exacerbations of skin lesions tend to parallel recurrences of the intestinal inflammation.[7]

The diagnosis of PG is based on the clinical appearance of the lesion, its association with systemic disease, the exclusion of other causes of dermatitis, and a poor response to antibiotics. The differential diagnosis of PG include bacterial infections, synergistic gangrene, deep fungal infection, necrotizing vasculitis, bullous erythema multiforme, Sweet syndrome, Behçet disease, halogen dermatitis, brown recluse spider bites, amebiasis, purpura fulminans, and factitial ulcer. The histopathological assessement of skin biopsy is necessary to rule out other diseases. Currently, specific laboratory or histopathological tests for PG are unavailable. In classic ulcerative PG, there is neutrophilic infiltrate centrally in the ulcer and lymphocytic infiltrate in the periphery.[5]

Management should be directed at both the lesions of PG and at the underlying disorder. Systemic corticosteroids are considered as the drug of choice for the treatment of PG. Prednisolone, 1 to 2 mg/kg/day are widely used for initial therapy. Pulse therapy with suprapharmacological doses of corticosteroids (500-1000 mg of methylprednisolone) is faster, but this treatment may cause fatal side effects in patients with cardiovascular diseases.[8,9] In many cases, cyclosporine is effective in the acute management of PG; however, when tapering the drug additional systemic agents are necessary for maintaining the clinical response.[8,10,11] It has recently been reported that TNF-alpha inhibitor infliximab is a safe and effective treatment in PG associated with inflammatory bowel disease.[12,13] Other systemic drugs that have been used , either alone or in conjunction with steroids are dapsone ,sulfasalazine, clofazimine, minocycline, potassium iodide, colchicines, human intravenous immune globulin, azathioprine, cyclophosphamide, chlorambucil, and tacrolimus.[14-20]

Moist wound management with an emphasis on preventing secondary infections is important. Topical sodium cromoglyate has been reported to be effective with and without systemic corticosteroid therapy.[20] The topical tacrolimus, hyperbaric oxygen also has been used successfully to treat PG.[21,22] Surgical debridement and/or skin grafting are not recommended during the acute stage, because of the high risk of pathergy.

In conclusion, systemic corticosteroids are the first-choice therapy in PG in spite of the presence of other many systemic and topical agents. Our patient's wounds healed with oral methyl prednisolon. The patient was therefore discharged with the recommendation of the steroid dose be tapered and finally stopped with 5-ASA treatment continuing under outpatient follow-up.

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