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Case Report

A CASE OF PYODERMA VEGETANS ASSOCIATED WITH CROHN'S DISEASE SHOWING GOOD RESPONSE TO PREDNISOLONE

Abstract

Pyoderma vegetans, also known as pyodermatitis vegetans, is a rare inflammatory dermatosis characterized by vesiculopustular lesions and vegetating plaques most commonly affecting the scalp, trunk, flexural areas and extremities. The etiopathogenesis of pyoderma vegetans has not been clearly elucidated yet; however, it has been proposed that pyoderma vegetans is associated with a number of underlying conditions, including ulcerative colitis, cutaneous T-cell lymphoma, primary immunodeficiency, alcoholism and human immunodeficiency virus infection. Although pyoderma vegetans may also be seen in healthy individuals, considerable association between immunosuppression and pyoderma vegetans is evident. Moreover, the occurrence of pyoderma vegetans and its oromucosal analogue, pyostomatitis vegetans, has been reported to be as high as 75% in patients with inflammatory bowel disease; hence, pyoderma vegetans has been considered to be an indicator of inflammatory bowel disease. On the other hand, the association of pyoderma vegetans with Crohn's disease has been rarely documented, and most of the reported cases are associated with ulcerative colitis. Here, we present a case of pyoderma vegetans associated with Crohn's disease that showed remarkable response to corticosteroid therapy.

Key Words: Pyoderma; Crohn Disease; Inflammatory Bowel Disease; Corticosteroids.

Olgu Sunumu

PREDNİZOLON TEDAVİSİNE İYİ YANIT VEREN CROHN HASTALIĞI İLE ASSOSİYE BİR PİYODERMA VEJETANS OLGUSU

Öz

Piyodermatitis vejetans olarak da adlandırılan piyoderma vejetans, genellikle saçlı deri, gövde, fleksural bölgeler ve ekstremitelerde yerleşen vezikülopüstüler lezyonlar ve vejetatif plaklarla karakterize nadir rastlanan inflamatuar bir dermatozdur. Piyoderma vejetans, etiyolojisi henüz tüm yönleriyle aydınlatılamamış sebebi bilinmeyen bir hastalıktır. Bununla birlikte, piyoderma vejetansın ülseratif kolit, kutanöz T hücreli lenfomalar, primer immün yetmezlikler, alkolizm ve insan immünyetmezlik virüsü gibi altta yatan çok sayıda durum ile bağlantılı olduğu ileri sürülmektedir. Piyoderma vejetans her ne kadar sağlıklı kişilerde de görülebilse de, piyoderma vejetans ile immün süpresyon arasındaki ilişki aşikardır. Dahası, piyoderma vejetans ve oromukozal analoğu piyostomatitis vejetansın, inflamatuar barsak hastalığı olan bireylerde %75'e varan oranlarda görüldüğü bildirilmiştir ki, piyoderma vejetansın inflamatuar barsak hastalığının bir göstergesi olduğu düşünülmektedir. Ancak, Crohn hastalığı ile assosiye olan piyoderma vejetans olgusu nadiren raporlanmıştır ve inflamatuar barsak hastalığı ile ilişkili olan piyoderma vejetans hastalarının çoğu ülseratif kolit tanısına sahiptir. Biz de burada, kortikosteroid tedavisine oldukça iyi yanıt veren Crohn hastalığı ile assosiye olan bir piyoderma vejetans olgusunu sunmak istiyoruz.

Anahtar Sözcükler: Piyoderma; Crohn Hastalığı; İnflamatuar Barsak Hastalığı; Kortikosteroidler.

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Introduction

Pyoderma vegetans (PV), or pyodermatitis vegetans, is a rare, chronic, inflammatory dermatosis characterized by vesiculopustular lesions and vegetating plaques most commonly affecting the scalp, flexural areas and extremities (1-3). When oral vegetative pustular oral lesions are present, it is called pyodermatitis-pyostomatitis vegetans that has been reported to be associated with inflammatory bowel disease (IBD) in 75% of cases (4). Here, we report a case of PV associated with Crohn's disease (CD) that showed good response to systemic prednisolone treatment.

Case Report

A 67-year-old male patient came to our outpatient clinic with a two-month history of non-healing wounds on his trunk and extremities. He had a medical history of asthma and CD, for which he was receiving inhaled glucocorticoids, long-acting bronchodilators and 40 mg/week of adalimumab. We were able to gather limited information regarding to his gastroenterological disease, in that the patient had been followed up for CD in another hospital. We learned that before the diagnosis of CD was made, he had been suffering from intractable abdominal pain, watery diarrhea and weight loss. He had been examined in a local hospital, where colonoscopy was performed, which revealed erythematous, friable colonic mucosa with skip areas. With the presumed diagnosis of CD, he was referred to a tertiary referral hospital which do not provide access to his medical records. However, we found out that one and a half years ago systemic corticosteroids and azathioprine were initiated for the diagnosis of CD although the treatment was changed with adalimumab one year ago. When he was admitted in our outpatient clinic, he was found to be anaemic with normal vital signs. On physical examination, the abdomen was soft and slightly tender on the right lower quadrant. Dermatological examination revealed multiple, vegetative, erythematous plaques with indurated borders studded with oozing pustules, particularly located on the pelvic, bilateral coxal and bilateral upper femoral areas. The peripheries of the plaques were serpiginous and the remaining central areas were hyperpigmented with velvety texture (Figure 1). Examination also revealed hyperpigmented, velvety plaques on the dorsum of the right hand, left popliteal area and several smaller erythematous plaques with central hyperpigmentation on the trunk. Laboratory tests demonstrated anaemia with a haemoglobin (Hb) level of 8.4 g/dL (normal range, 14–18 g/dL), serum iron level of 14 µg/dL (normal, 60–180 g/dL), transferrin saturation of 4% (normal, 20%–50%), serum ferritin level of 12.7 ng/mL (normal, 23.9–336.2 ng/mL), hypoalbuminemia with a serum albumin level of 2.5 g/dL (normal, 3.5–5.2 g/dL) and hypoproteinemia with serum a total protein level of 5.9 g/dL (normal, 6.5–8.5 g/dL). Moreover, the erythrocyte sedimentation rate (ESR) increased to 34 mm/h (normal, 0–20 mm/h), C-reactive protein level elevated to 62 mg/L (0,2–5 mg/L) and the faecal occult blood test was positive. Wound culture from one of the lesions was positive for *Staphylococcus aureus* and *Streptococcus* species. His-

Figure 1— Multiple erythematous, vegetating plaques of varying size with oozing pustules at the advancing serpiginous border and central velvety hyperpigmented areas on the pelvic, bilateral coxal and upper femoral regions.

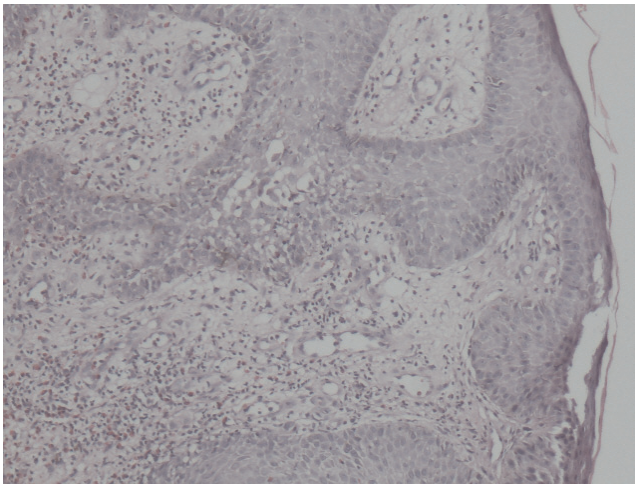

Figure 2— Focal suprabasal acantholysis is seen in epidermis that shows prominent pseudoepitheliomatous hyperplasia. There are many eosinophils in the dermis (HEx 100).

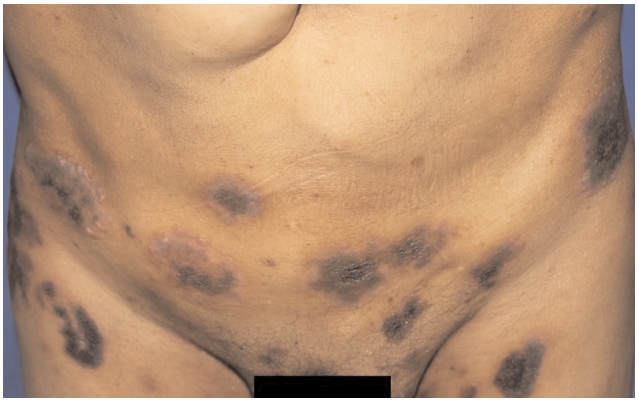

Figure 3— Resolution of the lesions after the theraphy (15th day of the treatment).

Figure 4— 45th day of the treatment.

topathological examination of the cutaneous lesions demonstrated pseudoepitheliomatous hyperplasia with dermal inflammatory infiltrate mainly comprising neutrophils, eosinophils and focal suprabasal acantholysis (Figure 2). Direct immunofluorescence studies were negative. The patient was assessed by the gastroenterologist however the consultant doctor did not planned any medical intervention due to the fact that the follow up of the patient had been carried out in another hospital. Based on clinical and histopathological findings, we diagnosed PV associated with CD. In addition to adalimumab, we administered oral prednisolone at 50 mg/day, topical corticosteroid and wet dressing with boric acid. On the 15th day visit, there was marked clinical improvement in both dermatological (Figure 3) and gastroenterological symptoms. As the patient showed clinical recovery (Figure 4), oral prednisolone was gradually tapered and discontinued within 3 months.

Discussion

Pyoderma vegetans is a rare chronic inflammatory dermatosis of uncertain aetiology characterized by vegetative oozing lesions and is generally observed in middle-aged men (1,3). However, PV has a high incidence rate in immunosuppressed states like immunoglobulin A deficiency (5), human immunodeficiency virus infection (6), diffuse T-cell lymphoma (7), chronic myeloid leukaemia (8), alcoholism and chronic malnutrition (2,3). On the other hand, although antibiotics usually offer limited improvement in PV symptoms, as *S. aureus* and other bacteria are generally isolated from PV lesions, bacterial colonization has been postulated as an aetiological factor for the induction of PV (1,2). Moreover, there have been reports regarding the considerable association between PV and halogen exposure, tattoos and foreign body reaction (2,3). Indeed, the most favoured hypothesis is that bacterial colonization or epidermal invasion plays a critical pathogenic role in the development of PV, which is believed to be an adverse tissue reaction, in patients with defective immunity who are locally or systemically immunocompromised (1,2).

To date, the highest prevalence of PV was found in patients with IBD, particularly ulcerative colitis (UC) (3,9-13). There is a general assumption that PV represents a specific marker for IBD (11) and a presumptive diagnosis of PV should prompt a complete gastrointestinal evaluation and necessitate the follow-up of the patient on a routine basis (2,3,13). Usually, the disease activity of PV runs a parallel course with that of IBD; PV progresses with intestinal disease, exacerbates with IBD and ameliorates with colectomy or remission of IBD (2,13). Systemic corticosteroids, dapsone, azathioprine, cyclosporine, isotretinoin, methotrexate and infliximab are treatment options for PV that generally cure both cutaneous and intestinal disease, as observed in our case (2,3,13-15). It is noteworthy that although the association of PV with CD has been rarely documented (4,16,17) and most of the reported cases are associated with UC (3,9-13), our case represents a particular example of PV associated with CD with characteristic clinical and histopathological findings, clinical course and response to treatment.

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