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Full Length Research paper

Scleroderma and pyoderma gangrenosum

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Pyoderma gangrenosum (PG) is an ulcerative disease of the skin with unknown origin. It may be associated with an underlying systemic disease like inflammatory bowel disease, lymphoproliferative disorders, sweet's syndrome and Behcet's disease. This case depicts a rare site of pyoderma gangronosum on scrotum. A 61-year-old man with scleroderma and pyoderma gangronosum on scrotum from two years ago. He had Skin thickening and hardening, hypertension, Raynaud's phenomenon, pulmonary hypertension, pulmonary fibrosis and multiple scrotal skin lesions.

Key words: Systemic sclerosis, pyoderma gangronosum.

INTRODUCTION

Pyoderma gangrenosum (PG) is an uncommon painful ulcerative skin condition which most often affects people in their 40 or 50 s. In 30% of cases, PG ulcerations may occur after trauma or injury to the skin. Rarely, PG may affect children and adolescents. It is associated with autoimmune diseases in at least 50% of patients such as inflammatory bowel disease, lymphoproliferative disorders, sweet's syndrome, Behcet's disease and scleroderma. The eruption may begin as an isolated pustule or scattered lesions on the trunk or extremities. Approximately one-half of cases are associated with an underlying systemic disease (Mir-Madjlessi et al., 1985; Powell et al., 1985; Hickman, 1983; Holt et al., 1980). PG has two main types and several other variants:

1. Classic PG is marked by a deep ulceration with a purple border that overhangs the ulcer bed. Lesions usually occur on the legs, but may occur elsewhere on the body. Classic PG near a stoma (the opening surgeons make after a colostomy) is often thought to be a wound infection or irritation.
2. Atypical PG has a pus- or fluid-filled component, typically at the border. It usually occurs on the back side of the hands, the forearms or the face. PG may occur on the genitalia (vulvar or penile PG). It is important to distinguish this variant from sexually transmitted diseases.

An intraoral (inside the mouth) form of the disease (pyostomatitis vegetans) occurs primarily in patients with inflammatory bowel disease. PG is sometimes found in the lungs, liver, and bones.

PG has also been described in patients with subcorneal pustulosis, and as a complication of therapy with G-CSF (Callen, 1989; Ross et al., 1991).

PG is usually observed on the legs; less typically on the hands. PG lesions can also occur in other organ systems, such as the heart, central nervous system, digestive tract, eyes, liver, spleen, bones and lymph nodes. PG generally responds well to treatment; however, it sometimes recurs or causes scarring (Callen, 1989).

The early lesions show mild to moderate perivascular mixed neutrophilic and lymphocytic infiltrate associated with endothelial swelling. Fully developed lesions consist of necrosis in addition to a dense neutrophilic infiltrate with some lymphocytes and macrophages surrounding and involving the blood vessels. Ulceration, infarction, and abscess formation are found in the later stages (Su et al., 1986).

CASE

We reported an unusual site of pyoderma gangrenosum on scrotum. A 61- year-old man complaining of lesions on the scrotum was referred to Al-Zahra Hospital. Patient two years ago with high blood pressure was treated with antihypertensive medications He had skin thickening and hardening, hypertension, Raynaud's phenomenon, pulmonary hypertension, and pulmonary fibrosis in the base of both lungs.

Drugs history include: Captopril 50 mg, Diltiazem 180

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Table 1. Laboratory data of our patient.

WBC: 16200/ l (4000-10,000)	LDH: 771 U/L (115-221)
Hb: 7.1g/dl (14-17)	ESR: 27 mm/h (3-15)
Plt: 299000/ l (150-450,000)	BUN: 64 mg/dl (8-24)
AST: 24 U/L (12-38)	Cr: 2.8 mg/dl (0.5-1.5)
ALT: 38 U/L (7-41)	U/A: normal
ALK-P: 178 IU/L (64- 306)	

Figure 1. Multiple skin lesions on scrotum.

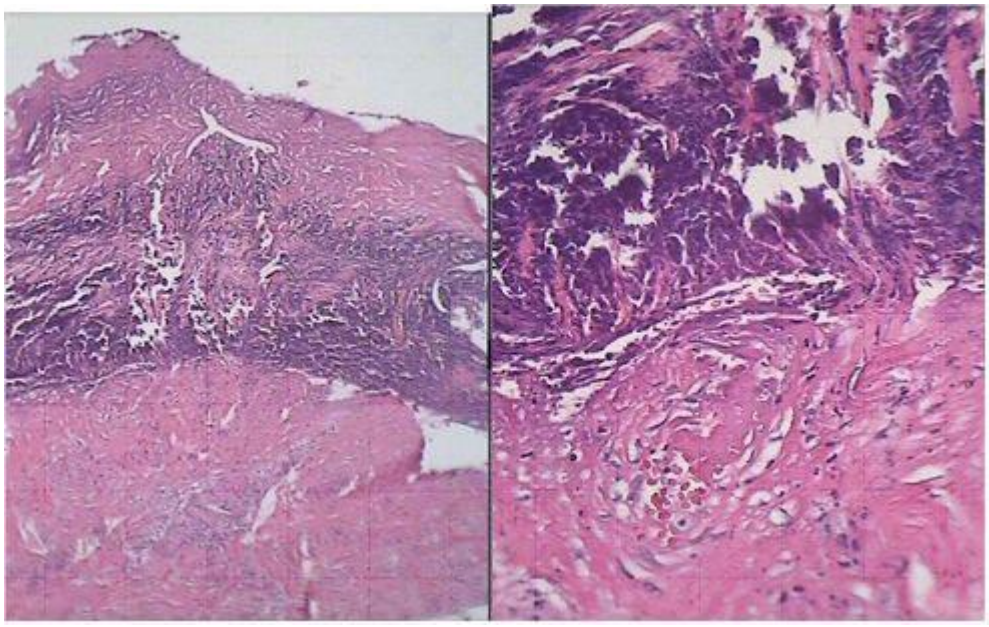

Figure 2. Necrosis of the epidermis and superficial dermis × 100.

mg, Losartan 100 mg, Prednisolone 5 mg, Sildenafil and Omeprazol 40 mg daily.

On physical examination there were multiple skin lesions on scrotum with erythematosus edges and necrosis with hemorrhage in the base of ulcers (Figure 1).

Ultrasonographic evaluation of both testis was normal except some degrees of skin thickness. The laboratory data were summarized in Table 1.

Skin biopsy from scrotum demonstrated some epidermal necrosis, hemorrhage, abscess formation, neutrophilic infiltration in epidermis and epidermal hyperplasia around the ulcer. By this data the diagnosis of pyoderma gangronosum was made (Figure 2).

The patient treated with prednisolone 0.5 mg / kg, oral azathioprine 1.5 mg / kg daily. In following up after 30 days the patient constitutional symptoms, general condition, and scrutum lesions was control. To prevent osteoporosis, calcium, vitamin D tablets to patients and weight-bearing exercise was prescribed.

DISCUSSION

Pyoderma gangrenosum is an inflammatory skin disease often associated with underlying systemic disorders such as inflammatory bowel disease, arthritis, and lympho-proliferative disorders. The eruption may begin as an isolated pustule or scattered lesions on the trunk or extremities. There is surrounding edema and purplish induration with rapid progression into a large ulcer which heals ultimately with cribiform scars. The diagnosis is typically made after all infectious etiologies have been ruled out. Histologic examination is helpful but not diagnostic in characterizing this disease.

Several reports along with pyoderma gangrenosum and autoimmune disorders have been reported in the following few are mentioned.

- Joshi and Mamta (2004) reported a case of behcets and pyoderma gangrenosum.
- Koskinas et al. (1999) reported a case of overlapping syndrome of autoimmune hepatitis and primary sclerosing cholangitis associated with pyoderma gangrenosum and ulcerative colitis.
- Stolman et al. (1975) reported two cases of pyoderma gangrenosum and rheumatoid arthritis.
- Pinto et al. (1991) reported pyoderma gangrenosum in a 35-year-old woman with long-standing systemic lupus erythematosus was treated with pulse steroid therapy.
- Vandevyvere et al. (2007) reported a case of pyoderma gangrenosum developing during therapy with TNF-alpha antagonists in a patient with rheumatoid arthritis.
- Yayli et al. (2005) reported a 17-year-old girl with JRA presented with pyoderma gangrenosum.- Fujikura et al. association with peristomal pyoderma gangrenosum. Pyoderma gangrenosum occurs in association not only with inflammatory bowel disease, but also with other diseases (Fujikura et al., 2007).

There is a case report of limited scleroderma with ostomal pyoderma gangronosum by Fujikura et al. (2007)

Treatment options

Pyoderma gangrenosum can be treated effectively, but complete healing can take months. Small ulcers are usually treated with topical creams and dressings, while large ulcers require immunosuppressive therapy (Corticosteroids, Cyclosporine, Azathioprine, Tacrolimus, Mycophenolatemofetil, Dapsone, Cyclophosphamide pulse therapy[1], Thalidomide) in sever cases Biological therapies (anti-tumour necrosis factor treatments) are effective (Brooklyn et al., 2006; Rogge et al., 2008).

Other treatments reported as helpful (in case reports) a are: Low dose colchicine, Minocycline, Intravenous immu noglobulin, Clofazimine, Leukophoresis, Hyperbaric oxyg en therapy (Wollina et al., 2007).

Surgical procedures are rarely used because they can aggravate the condition. Our patient was treated with Prednisolone 15 mg and azathioprine 150 mg daily; 30 days after beginning of drugs all of lesions disappeared.

CONCLUSION

In conclusion, we report a rare case of scleroderma in an adult man with scrutum pyoderma gangrenosum. The patient treated with prednisolone (0.5 mg/kg) and oral azathioprine (1.5 mg/kg daily).

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