

# World Journal of *Clinical Cases*

World J Clin Cases 2018 November 6; 6(13): 577-715





### REVIEW

- 577 Role of bile acids in colon carcinogenesis  
*Nguyen TT, Ung TT, Kim NH, Jung YD*

### MINIREVIEWS

- 589 Update on global epidemiology of viral hepatitis and preventive strategies  
*Jefferies M, Rauff B, Rashid H, Lam T, Rafiq S*

### ORIGINAL ARTICLE

#### Case Control Study

- 600 Iron metabolism disorders in patients with hepatitis B-related liver diseases  
*Gao YH, Wang JY, Liu PY, Sun J, Wang XM, Wu RH, He XT, Tu ZK, Wang CG, Xu HQ, Niu JQ*

#### Retrospective Cohort Study

- 611 Impact of an acute hemodynamic response-guided protocol for primary prophylaxis of variceal bleeding  
*Forteza JI, Puente Á, Ruiz P, Ezcurra I, Vaquero J, Cuadrado A, Arias-Loste MT, Cabezas J, Llerena S, Iruzubieta P, Rodríguez-Lope C, Huelin P, Casafont F, Fábrega E, Crespo J*

#### Retrospective Study

- 624 Effect of a region-wide incorporation of an algorithm based on the 2012 international consensus guideline on the practice pattern for the management of pancreatic cystic neoplasms in an integrated health system  
*Nguyen AK, Girg A, Tekeste T, Chang K, Adeyemo M, Eskandari A, Alonso E, Yaramada P, Chaya C, Ko A, Burke E, Roggow I, Butler R, Kawatkar A, Lim BS*

- 632 Usefulness of colonic tattooing using indocyanine green in patients with colorectal tumors  
*Park JH, Moon HS, Kwon IS, Yun GY, Lee SH, Park DH, Kim JS, Kang SH, Lee ES, Kim SH, Sung JK, Lee BS, Jeong HY*

#### Randomized Clinical Trial

- 641 *Helicobacter pylori* may be an initiating factor in newly diagnosed ulcerative colitis patients: A pilot study  
*Mansour L, El-Kalla F, Kobtan A, Abd-Elsalam S, Yousef M, Soliman S, Ali LA, Elkhawany W, Amer I, Harras H, Hagar MM, Elhendawy M*

### META-ANALYSIS

- 650 Photodynamic therapy for middle-advanced stage upper gastrointestinal carcinomas: A systematic review and meta-analysis  
*Chen B, Xiong L, Chen WD, Zhao XH, He J, Zheng YW, Kong FH, Liu X, Zhang ZJ, Miao XY*

### CASE REPORT

- 659** Successful rescue of acute liver failure and hemophagocytic lymphohistiocytosis following varicella infection: A case report and review of literature  
*Zhang LN, Guo W, Zhu JH, Guo Y*
- 666** Bilateral thoracic kidneys combined with inferior vena cava located behind the anterior abdominal wall: A case report and review of literature  
*Peng XX, Cheng SA, Liang QL, Luo XB, Hong XC, Yuan GL, Zhang HJ*
- 671** Incident hepatocellular carcinoma developing during tenofovir alafenamide treatment as a rescue therapy for multi-drug resistant hepatitis B virus infection: A case report and review of the literature  
*Lu JC, Liu LG, Lin L, Zheng SQ, Xue Y*
- 675** Possible connection between elevated serum  $\alpha$ -fetoprotein and placental necrosis during pregnancy: A case report and review of literature  
*Yu MY, Xi L, Zhang JX, Zhang SC*
- 679** Laparoscopic pancreatic duct incision and stone removal and T-type tube drainage for pancreatic duct stone: A case report and review of literature  
*Bai Y, Yu SA, Wang LY, Gong DJ*
- 683** Detection of a unicentric type of Castleman-like mass at the site of adrenal gland: A case report and review of literature  
*Chen J, Yang C, Liang CZ*
- 688** Systemic lupus erythematosus complicated by noncirrhotic portal hypertension: A case report and review of literature  
*Yang QB, He YL, Peng CM, Qing YF, He Q, Zhou JG*
- 694** Natural killer/T-cell lymphoma with concomitant syndrome of inappropriate antidiuretic hormone secretion: A case report and review of literature  
*Liu QB, Zheng R*
- 703** Successful treatment of pyoderma gangrenosum with concomitant immunoglobulin A nephropathy: A case report and review of literature  
*Li XL, Ma ZG, Huang WH, Chai EQ, Hao YF*



- 707 Highlighting the importance of early diagnosis in progressive multi-organ involvement of IgG4-related disease: A case report and review of literature

*Xue J, Wang XM, Li Y, Zhu L, Liu XM, Chen J, Chi SH*

**ABOUT COVER**

Editorial Board Member of *World Journal of Clinical Cases*, Byung-Wook Kim, MD, PhD, Professor, Division of Gastroenterology, Department of Internal Medicine, Incheon St. Mary's Hospital, the Catholic University of Korea, Incheon 21431, South Korea

**AIM AND SCOPE**

*World Journal of Clinical Cases* (*World J Clin Cases*, *WJCC*, online ISSN 2307-8960, DOI: 10.12998) is a peer-reviewed open access academic journal that aims to guide clinical practice and improve diagnostic and therapeutic skills of clinicians.

The primary task of *WJCC* is to rapidly publish high-quality Autobiography, Case Report, Clinical Case Conference (Clinicopathological Conference), Clinical Management, Diagnostic Advances, Editorial, Field of Vision, Frontier, Medical Ethics, Original Articles, Clinical Practice, Meta-Analysis, Minireviews, Review, Therapeutics Advances, and Topic Highlight, in the fields of allergy, anesthesiology, cardiac medicine, clinical genetics, clinical neurology, critical care, dentistry, dermatology, emergency medicine, endocrinology, family medicine, gastroenterology and hepatology, geriatrics and gerontology, hematology, immunology, infectious diseases, internal medicine, obstetrics and gynecology, oncology, ophthalmology, orthopedics, otolaryngology, pathology, pediatrics, peripheral vascular disease, psychiatry, radiology, rehabilitation, respiratory medicine, rheumatology, surgery, toxicology, transplantation, and urology and nephrology.

**INDEXING/ABSTRACTING**

*World Journal of Clinical Cases* (*WJCC*) is now indexed in PubMed, PubMed Central, Science Citation Index Expanded (also known as SciSearch®), and Journal Citation Reports/Science Edition. The 2018 Edition of Journal Citation Reports cites the 2017 impact factor for *WJCC* as 1.931 (5-year impact factor: N/A), ranking *WJCC* as 60 among 154 journals in Medicine, General and Internal (quartile in category Q2).

**EDITORS FOR THIS ISSUE**

**Responsible Assistant Editor:** *Xiang Li*  
**Responsible Electronic Editor:** *Yun-XiaoJian Wu*  
**Proofing Editor-in-Chief:** *Lian-Sheng Ma*

**Responsible Science Editor:** *Ying Dou*  
**Proofing Editorial Office Director:** *Jin-Lei Wang*

**NAME OF JOURNAL**  
*World Journal of Clinical Cases*

**ISSN**  
ISSN 2307-8960 (online)

**LAUNCH DATE**  
April 16, 2013

**FREQUENCY**  
Semimonthly

**EDITORS-IN-CHIEF**  
**Sandro Vento, MD**, Department of Internal Medicine, University of Botswana, Private Bag 00713, Gaborone, Botswana

**EDITORIAL BOARD MEMBERS**  
All editorial board members resources online at <http://www.wjgnet.com/2307-8960/editorialboard.htm>

**EDITORIAL OFFICE**  
Jin-Lei Wang, Director

*World Journal of Clinical Cases*  
Baishideng Publishing Group Inc  
7901 Stoneridge Drive, Suite 501, Pleasanton, CA 94588, USA  
Telephone: +1-925-2238242  
Fax: +1-925-2238243  
E-mail: [editorialoffice@wjgnet.com](mailto:editorialoffice@wjgnet.com)  
Help Desk: <http://www.f6publishing.com/helpdesk>  
<http://www.wjgnet.com>

**PUBLISHER**  
Baishideng Publishing Group Inc  
7901 Stoneridge Drive, Suite 501, Pleasanton, CA 94588, USA  
Telephone: +1-925-2238242  
Fax: +1-925-2238243  
E-mail: [bpgoffice@wjgnet.com](mailto:bpgoffice@wjgnet.com)  
Help Desk: <http://www.f6publishing.com/helpdesk>  
<http://www.wjgnet.com>

**PUBLICATION DATE**  
November 6, 2018

**COPYRIGHT**

© 2018 Baishideng Publishing Group Inc. Articles published by this Open Access journal are distributed under the terms of the Creative Commons Attribution Non-commercial License, which permits use, distribution, and reproduction in any medium, provided the original work is properly cited, the use is non commercial and is otherwise in compliance with the license.

**SPECIAL STATEMENT**

All articles published in journals owned by the Baishideng Publishing Group (BPG) represent the views and opinions of their authors, and not the views, opinions or policies of the BPG, except where otherwise explicitly indicated.

**INSTRUCTIONS TO AUTHORS**

<http://www.wjgnet.com/bpg/gerinfo/204>

**ONLINE SUBMISSION**

<http://www.f6publishing.com>

## Successful treatment of pyoderma gangrenosum with concomitant immunoglobulin A nephropathy: A case report and review of literature

Xiao-Li Li, Zhi-Gang Ma, Wen-Hui Huang, Er-Qing Chai, Yun-Fei Hao

Xiao-Li Li, Zhi-Gang Ma, Wen-Hui Huang, Department of Nephrology, Gansu Provincial Hospital, Lanzhou 730000, Gansu Province, China

Yun-Fei Hao, Er-Qing Chai, Cerebrovascular Disease Center, Gansu Provincial Hospital, Lanzhou 730000, Gansu Province, China

ORCID number: Xiao-Li Li (0000-0002-9821-5434); Zhi-Gang Ma (0000-0002-5223-2976); Wen-Hui Huang (0000-0002-8776-8834); Er-Qing Chai (0000-0001-5251-4626); Yun-Fei Hao (0000-0003-1441-3722).

**Author contributions:** Hao YF and Ma ZG designed the study. Li XL and Huang WH collected the patient's clinical data. Chai EQ analyzed the data and wrote the paper.

**Informed consent statement:** The patient involved in this study gave her written informed consent authorizing the use and disclosure of her protected health information.

**Conflict-of-interest statement:** No potential conflicts of interest relevant to this article were reported.

**CARE Checklist (2013) statement:** The authors have read the CARE Checklist (2013), and the manuscript was prepared and revised according to the CARE Checklist (2013).

**Open-Access:** This article is an open-access article which was selected by an in-house editor and fully peer-reviewed by external reviewers. It is distributed in accordance with the Creative Commons Attribution Non Commercial (CC BY-NC 4.0) license, which permits others to distribute, remix, adapt, build upon this work non-commercially, and license their derivative works on different terms, provided the original work is properly cited and the use is non-commercial. See: <http://creativecommons.org/licenses/by-nc/4.0/>

**Manuscript source:** Unsolicited manuscript

**Correspondence to:** Yun-Fei Hao, MD, Attending Doctor, Cerebrovascular Disease Center, Gansu Provincial Hospital, No. 204, Donggang West Road, Lanzhou 730000, Gansu Province,

China. [hyf897@sina.com](mailto:hyf897@sina.com)

Telephone: +86-18893114576

Received: July 30, 2018

Peer-review started: July 30, 2018

First decision: August 31, 2018

Revised: September 20, 2018

Accepted: October 11, 2018

Article in press: October 12, 2018

Published online: November 6, 2018

### Abstract

Pyoderma gangrenosum (PG) is an uncommon ulcerative cutaneous condition of an unknown etiology and is often associated with immune diseases. However, PG rarely shows visceral involvement, especially in the kidney. A 20-year-old female presented with pedal edema and skin ulceration of both lower limbs. The skin lesion began as an erythematous plaque and then became a blister. She also complained of abdominal distension and a decreasing urine volume. Laboratory data showed high proteinuria, hypoalbuminemia and hyperlipidemia. Her skin and kidney were biopsied. The pathological results indicated PG and immunoglobulin A (IgA) nephropathy. The patient was finally cured with prednisolone in combination with cyclosporine A (CsA).

**Key words:** Pyoderma gangrenosum; Immunoglobulin A nephropathy; Treatment

© The Author(s) 2018. Published by Baishideng Publishing Group Inc. All rights reserved.

**Core tip:** This is the first report of successfully treated pyoderma gangrenosum (PG) occurring concurrently with immunoglobulin A (IgA) nephropathy. Both are immune-mediated disorders and should be paid attention to.



Li XL, Ma ZG, Huang WH, Chai EQ, Hao YF. Successful treatment of pyoderma gangrenosum with concomitant immunoglobulin A nephropathy: A case report and review of literature. *World J Clin Cases* 2018; 6(13): 703-706 Available from: URL: <http://www.wjgnet.com/2307-8960/full/v6/i13/703.htm> DOI: <http://dx.doi.org/10.12998/wjcc.v6.i13.703>

## INTRODUCTION

Pyoderma gangrenosum (PG) is an uncommon, ulcerative, cutaneous condition of an unknown cause, with an estimated annual incidence of 3-10 cases per million in the population<sup>[1]</sup>. PG is usually associated with systemic diseases such as inflammatory bowel disease, rheumatoid arthritis, seronegative arthritis, and autoimmune hepatitis and hematologic disorders such as paraproteinemia (especially immunoglobulin A paraproteinemia) and neutrophil malignancies<sup>[2]</sup>, most of which exhibit mucocutaneous involvement. PG with visceral (especially renal) involvement is rare. Here, we report, to the best of our knowledge, the first case of a patient with PG in combination with immunoglobulin A (IgA) nephropathy, who was successfully treated with a glucocorticoid in combination with cyclosporine A (CsA).

## CASE REPORT

A 20-year-old female presented with swelling and ulceration of both lower limbs, which lasted for 1 wk. The skin lesion began as an erythematous plaque and then became a blister. In spite of antibiotic treatment and wound care, the lesion progressed for 1 wk as a painful ulceration of 3-5 cm in diameter, with a violaceous border and purulent or sanguineous exudate at the base (Figure 1). Additionally, she reported mucopurulent bloody stool and severe abdominal heaviness, but no fever, weight loss, arthralgia or other signs or symptoms of systemic illness.

The laboratory workup revealed moderate anemia (87 g/L), slightly increased C-reactive protein (33.8 mg/L) and ESR (27 mmol/L) levels, and negativity for autoantibodies, rheumatoid factor and antistreptolysin O. Additionally, high proteinuria (13 g/24 h), hypoalbuminemia (16 g/L) and hyperlipidemia were observed. Stool tests showed pyocytes and red blood cells, but no bacterial cultures were obtained. Abdominal ultrasound indicated massive ascites. The skin lesions were cultured for bacteria and *Mycobacterium tuberculosis*, but the results were negative.

The edges of the lesions were biopsied. The histological results showed massive small lymphocytes arranged around blood vessels throughout the dermis (Figure 2). In addition, renal biopsy was performed. Light microscopy showed moderate enlargement of the mesangial area caused by an increase mesangial cells and the matrix as well as diffuse proliferation and degeneration of endothelial cells, infiltrated with neutrophils (Figure 3).

Immunofluorescence analysis showed deposition of IgA and complement 3 in the mesangial area (Figure 4).

Prednisolone at 1 mg/kg plus cyclophosphamide at 0.6 mg/2 wk were prescribed. After 2 wk, the stool had returned to normal, and the skin lesions had improved. However, proteinuria, oliguria, and ascites were not alleviated after 2 mo of treatment. Thereafter, prednisolone was tapered off at 10% of the dosage every 10 d until the dose reached 5 mg, and cyclophosphamide was replaced by CsA 3 mg/(kg·d) (75 mg *b.i.d.*). After 2 wk, urine output increased to normal. Other renal symptoms were also gradually alleviated. In month 4, urine protein disappeared, and CsA was then tapered off at 25 mg every 2 mo. One year later, all indices were normal, with only pigmentation remaining in the skin lesions (Figure 5).

## DISCUSSION

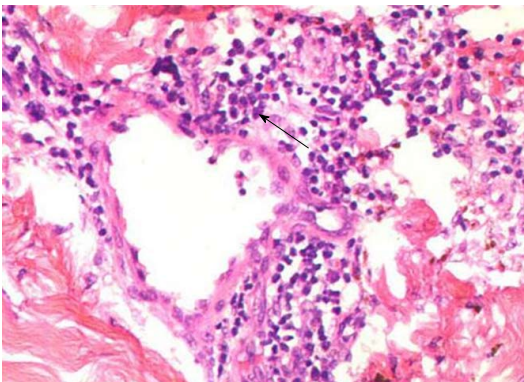
PG was first described by Brocq in 1916 and further characterized by Perry *et al.*<sup>[3]</sup>. It can affect an individual at any age but usually occurs between the ages of 20 and 50 years, with female predominance<sup>[4,5]</sup>. Skin lesions typically appear on the lower limbs but may also be observed on the upper extremities, head, and neck and even the genitals. It is diagnosed clinically, with no specific laboratory tests. The diagnosis is mainly based on the criteria proposed by Su *et al.*<sup>[6]</sup> in 2004, including two major criteria: (1) rapid progression of a painful, necrotic cutaneous ulcer with an irregular, violaceous, and undermined border, and (2) exclusion of other causes of cutaneous ulceration; at least two minor criteria must also be present: (1) a history suggestive of pathergy or a clinical finding of cribriform scarring, (2) concomitant systemic diseases, (3) histopathologic findings of sterile dermal neutrophilia,  $\pm$  mixed inflammation  $\pm$  lymphocytic vasculitis, and (4) a rapid response to systemic steroid treatment<sup>[6]</sup>. Of note, the histopathologic findings are not specific and may vary with the biopsy site and duration of the disease<sup>[7]</sup>. Thus, the diagnosis of PG is established based on characteristic clinical features, a good response to treatment and exclusion of infections (by bacteria, fungi, and typical or atypical mycobacteria), neoplastic disorders, and vasculitic disorders by biopsy and culture.

The etiology of PG remains unknown. However, 50% of cases are associated with systemic diseases, which are most commonly autoimmune disorders, suggesting that dysregulation of the immune system plays a role in disease pathogenesis. IgA nephropathy also involves immune-mediated inflammation of the glomeruli of the kidney and is characterized by deposition of the IgA antibody in the glomerular mesangium. However, PG and IgA nephropathy have not previously been concomitantly found; this is the first case report involving these two diseases.

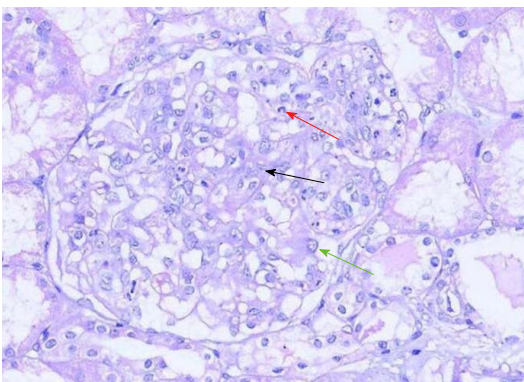
The first-line modality for both diseases is systemic corticosteroids. The mainstay of treatment is long-term immunosuppression, often with a high dose of corticosteroids [prednisolone 0.5-2 mg/(kg·d)] or a low dose



**Figure 1** The right lower leg exhibited ulcerated lesions with erythematous-violaceous excavated borders and a necrotic center.

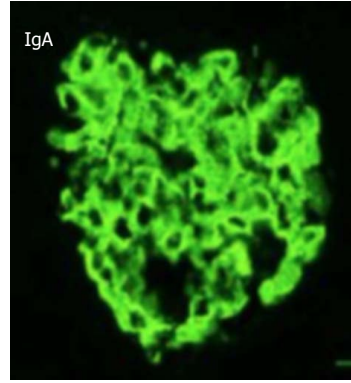


**Figure 2** Light microscopy of the primary skin lesion. Massive small lymphocytes (black arrow) are arranged around blood vessels throughout the dermis [hematoxylin and eosin (HE) staining,  $\times 200$ ]



**Figure 3** Light microscopy of the biopsied kidney tissue. The mesangial area is moderately enlarged due to an increase in mesangial cells (black arrow) and the matrix. Endothelial cells (green arrow) show diffuse proliferation and degeneration. Infiltrated neutrophils (red arrow) are present [hematoxylin and eosin (HE) staining,  $\times 200$ ].

of CsA [3-6 mg/(kg·d)]. In the present case, we initially prescribed prednisolone at 1 mg/kg daily in combination with cyclophosphamide at 0.6 mg/2 wk. However, the kidneys and skin did not show a parallel response. The skin lesion improved rapidly, whereas the kidney-related symptoms showed no remission until cyclophosphamide was replaced by CsA.



**Figure 4** Immunofluorescence staining of the biopsied kidney tissues. IgA showed strong positivity within the mesangium ( $\times 200$ ). IgA: Immunoglobulin A.



**Figure 5** The skin lesions on the right lower leg were healed after one year, with only pigmentation remaining.

The possible immunological link between the two disease entities remains unclear. Physicians should always bear in mind the possibility of a diagnosis of IgA nephropathy in PG patients. We also wish to present this unusual case in the hope that it will provide a valuable contribution to the treatment of the disease and to the literature.

To our knowledge, this is the first report of successfully treated PG occurring concurrently with IgA nephropathy. Both are immune-mediated disorders and can be cured with prednisolone in combination with CsA.

## ARTICLE HIGHLIGHTS

### Case characteristics

A 20-year-old female with pyoderma gangrenosum (PG) with concomitant immunoglobulin A (IgA) nephropathy was successfully treated.

### Clinical diagnosis

According to the laboratory results and clinical manifestations, nephric syndrome and PG were diagnosed.

### Differential diagnosis

Skin infections should be excluded.

### Laboratory diagnosis

High proteinuria (13 g/24 h), hypoalbuminemia (16 g/L) and hyperlipidemia



suggested nephrotic syndrome.

### Imaging diagnosis

Abdominal ultrasound indicated normally sized kidneys and massive ascites.

### Pathological diagnosis

Renal biopsy showed IgA nephropathy, with stronger staining (3+) for IgA and C3 in the mesangial area, and skin biopsy indicated massive small lymphocytes arranged around blood vessels throughout the dermis, suggesting vasculitis.

### Treatment

Prednisolone at 1 mg/kg plus cyclophosphamide at 0.6 mg/2wk, followed by prednisone at 5 mg/kg plus cyclosporine A (CsA) at 3 mg/(kg·d) (75 mg *b.i.d.*).

### Related reports

There are no reports of coexistence of PG and IgA nephropathy. This is the first reported case of PG with concomitant IgA nephropathy to be successfully treated.

### Term explanation

PG and IgA nephropathy are both autoimmune diseases.

### Experiences and lessons

Low-dosage prednisone (5 mg/kg) plus CsA may be helpful in patients with PG concomitant with IgA nephropathy.

## ACKNOWLEDGMENTS

We wish to thank Dr. Ma for his help in the diagnosis

and treatment of the patient.

## REFERENCES

- 1 **Ahronowitz I**, Harp J, Shinkai K. Etiology and management of pyoderma gangrenosum: a comprehensive review. *Am J Clin Dermatol* 2012; **13**: 191-211 [PMID: 22356259 DOI: 10.2165/11595240-000000000-00000]
- 2 **Alavi A**, French LE, Davis MD, Brassard A, Kirsner RS. Pyoderma Gangrenosum: An Update on Pathophysiology, Diagnosis and Treatment. *Am J Clin Dermatol* 2017; **18**: 355-372 [PMID: 28224502 DOI: 10.1007/s40257-017-0251-7]
- 3 **Perry HO**, Brunsting LA. Pyoderma gangrenosum; a clinical study of nineteen cases. *AMA Arch Derm* 1957; **75**: 380-386 [PMID: 13402210 DOI: 10.1001/archderm.1957.01550150066007]
- 4 **Gameiro A**, Pereira N, Cardoso JC, Gonçalo M. Pyoderma gangrenosum: challenges and solutions. *Clin Cosmet Investig Dermatol* 2015; **8**: 285-293 [PMID: 26060412 DOI: 10.2147/CCID.S61202]
- 5 **Brown TS**, Marshall GS, Callen JP. Cavitating pulmonary infiltrate in an adolescent with pyoderma gangrenosum: a rarely recognized extracutaneous manifestation of a neutrophilic dermatosis. *J Am Acad Dermatol* 2000; **43**: 108-112 [PMID: 10863234 DOI: 10.1067/mjd.2000.103627]
- 6 **Su WP**, Davis MD, Weenig RH, Powell FC, Perry HO. Pyoderma gangrenosum: clinicopathologic correlation and proposed diagnostic criteria. *Int J Dermatol* 2004; **43**: 790-800 [PMID: 15533059 DOI: 10.1111/j.1365-4632.2004.02128.x]
- 7 **Callen JP**, Jackson JM. Pyoderma gangrenosum: an update. *Rheum Dis Clin North Am* 2007; **33**: 787-802, vi [PMID: 18037117 DOI: 10.1016/j.rdc.2007.07.016]

**P- Reviewer:** Choi MR, Ekpenyong CEE, Nechifor G  
**S- Editor:** Ma RY **L- Editor:** A **E- Editor:** Wu YXJ





Published by **Baishideng Publishing Group Inc**  
7901 Stoneridge Drive, Suite 501, Pleasanton, CA 94588, USA  
Telephone: +1-925-223-8242  
Fax: +1-925-223-8243  
E-mail: [bpgoffice@wjgnet.com](mailto:bpgoffice@wjgnet.com)  
Help Desk: <http://www.f6publishing.com/helpdesk>  
<http://www.wjgnet.com>

