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CASE REPORT

Peristomal pyoderma gangrenosum successfully treated with cyclosporine

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ABSTRACT

Peristomal pyoderma gangrenosum (PPG) is an uncommon subtype of pyoderma gangrenosum. We describe a 74-year-old woman without inflammatory bowel disease (IBD) who developed PG around the jejunostomy and was successfully treated with cyclosporine. In our case, irritations by a lumbar corset and repeated sutures around the jejunostomy seemed to have caused pathergy reaction. It is noteworthy that 20% of PPG patients do not have IBD and therefore patients presenting PPG without IBD should not be overlooked.

Keywords: Peristomal Pyoderma Gangrenosum; Jejunostomy; Pathergy; Cyclosporine

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1. Introduction

A pyoderma gangrenosum (PG) subtype, peristomal pyoderma gangrenosum (PPG), occurs after surgical placement of an ileostomy or colostomy, usually due to underlying inflammatory bowel disease (IBD)[1,2]. Twenty percent of patients with PPG, however, had ostomies placed for other causes than IBD, including gastro-intestinal cancer, bladder cancer, bowel perforation and posterior urethral valves[3]. It is very rare that PPG develops around the jejunostomy or gastrostomy because of the differences in underlying diseases, irritation from ostomy leakage and frequency in appliance changes[4]. Effective PPG management requires either local or systemic immunosuppression and ostomy-compatible wound cares. We herein report a case of PPG developed around jejunostomy and it was successfully treated with cyclosporine.

2. Case presentation

In this case we describe the management of PPG occurring at a jejunostomy site in a 74-year-old woman with a lower esophageal cancer. Based on the findings of blood examination and colonoscopy, we denied the coexistence of hematological malignancy, rheumatic arthritis, inflammatory bowel disease and autoimmune disorders.

The patient underwent a subtotal esophagectomy, gastric tube reconstruction and jejunostomy. Three weeks later, she fell down and broke the third lumbar vertebra. She had to wear a lumbar corset. Two months after the operation, the redness and exudates appeared

around the jejunostomy. The jejunostomy tube was repeatedly sutured to connect to the peristomal skin because of the loosing of the ligation. The redness around the ostomy rapidly progressed into a painful ulcer with serpiginous and violaceous borders with surrounding erythema (**Figure 1A**). Topical use of corticosteroid was not effective. Skin biopsy specimens obtained from the ulcer edge demonstrated massive neutrophilic infiltrate. Swabs of the ulcerated skin revealed the commensal skin and gut flora. The typical clinical features such as rapidly expanding peristomal ulcers with pathergy and neutrophilic dermal infiltrates induced the diagnosis of PPG. She stopped wearing the lumbar corset. Cyclosporine 100 mg daily was started and the ulcer was completely healed one month later. Then cyclosporine was tapered and stopped within three months without any recurrence (**Figure 1B**).

3. Discussion

Peristomal pyoderma gangrenosum (PPG) is an uncommon subtype of pyoderma gangrenosum and the etiology of PPG remains poorly understood. Regarding the diagnosis, we can apply the PARACELSUS score which is a diagnostic tool for pyoderma gangrenosum[5]. According to this scoring system for PG, our patient showed 19 points, which indicates a highly likelihood of PG. Based on the systematic review[3], an 81% of PPG patients had a diagnosis of IBD and a 19% had ostomies placed for indications other than IBD. Although immunologic abnormalities may be important in its pathophysiology, risk factors for recurrent pathergy including ostomy leakage, appliance changes and tension or ischemia by ostomy appliances are also strongly related to the appearance of PPG.

Our patient had a jejunostomy due to a subtotal esophagectomy without IBD. PPG is very rare in jejunostomy because the risk factors are less than PPG involving ileostomy or colostomy. In our case, we strongly suspect that both mechanical irritations by a lumbar corset and repeated sutures caused the pathergy reaction.

PPG management entails controlling inflammation and optimizing wound care.

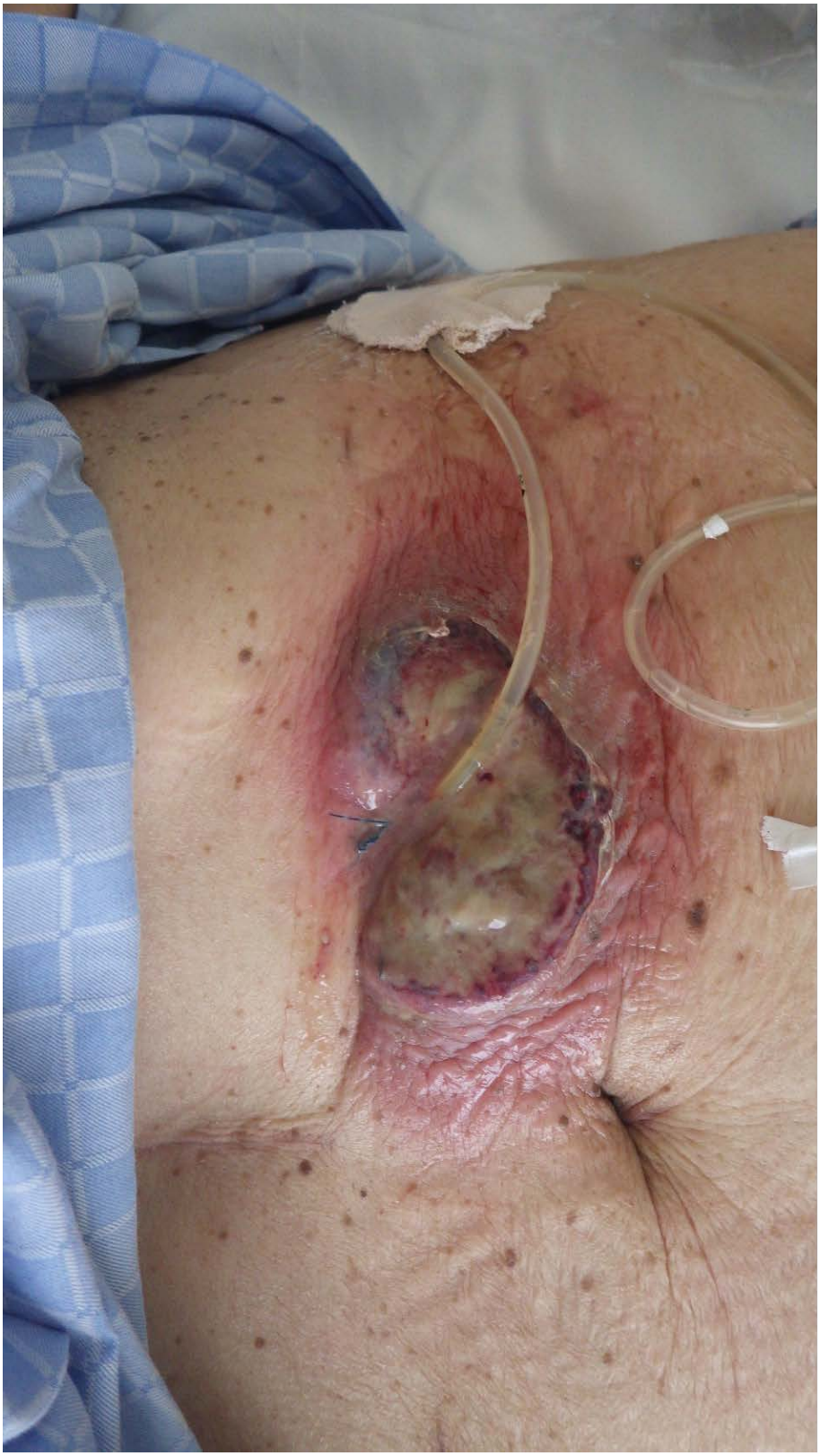
Efficacies of PPG therapies and recommendations by previous studies are summarized by Afifi *et al.*[3], based on Oxford Centre for Evidence-Based Medicine grade of recommendation. Mild cases of PPG without active systemic disease can be managed with wound care and topical agents. More severe cases or those with systemic disease may require a systemic medication or surgical interventions. The overall goals of wound care are to provide a clean wound environment, absorb exudate, maintain moderate moisture, and prevent further skin irritation. Topical therapies for PPG treatment include corticosteroids and calcineurin inhibitors with similar clinical responsiveness. Systemic therapies are important as the first-line treatments for severe or rapidly evolving cases of PPG. Grade C systemic immunosuppressive therapies including corticosteroids, cyclosporine (3–5 mg/kg/day) and dapsone (100 mg) showed complete responses in 50% of patients. Although corticosteroids were the most commonly used systemic treatment for PPG, similar effectiveness has been found by systemic cyclosporine. Systemic metronidazole, azathioprine, sulfasalazine and tacrolimus were less commonly used and can be considered effective alternatives or adjunct therapies. The biologic agents such as infliximab and adalimumab have yielded beneficial results and are considered grade C recommendations.

In our case, cyclosporine (2 mg/kg/day) was greatly effective and the ulcer didn't appear again even after its withdrawal of cyclosporin. During the cyclosporine treatment, the patient didn't show the adverse drug reactions such as hypertension and renal dysfunction. Since she experienced the third lumbar vertebra fractured just before the occurrence of PPG, we gave a priority to a cyclosporine treatment over corticosteroid.

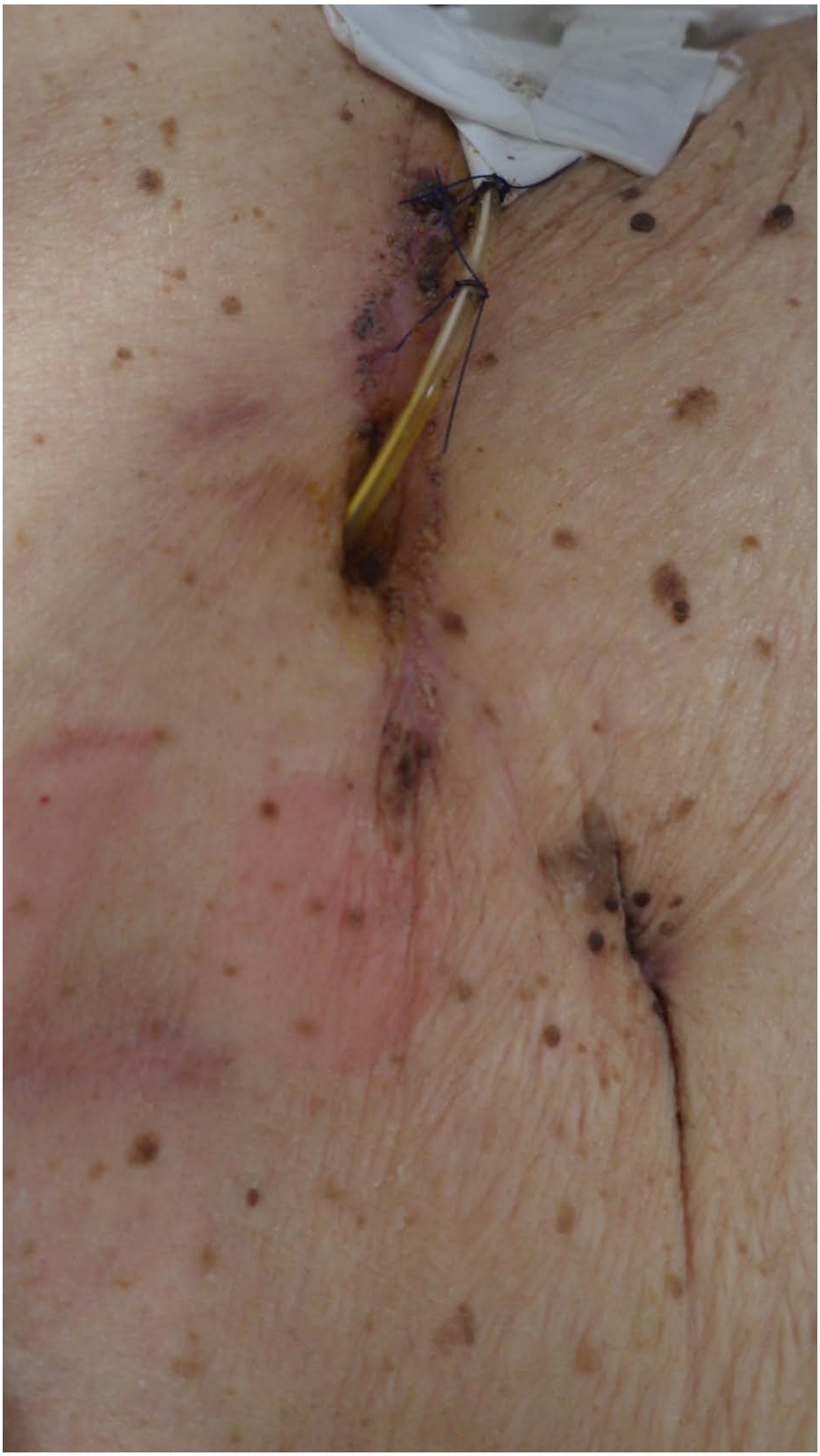
It is also noteworthy that cyclosporin was demonstrated useful for initial and/or maintenance therapy as the practical treatment for intractable inflammatory diseases[6,7].

It should be noticed that PPG can occur in a patient without IBD even around a jejunostomy or gastrostomy. For patients with peristomal ulcer, risk factors of pathergy should be minimized and therapeutic decision should be made as soon as possi-

ble based on the accurate diagnosis.



(A)



(B)

Figure 1. Skin features of PPG before (**A**) and after the treatment by cyclosporine (**B**).

4. Conclusion

This patient of PPG which developed around jejunostomy was successfully treated by minimizing the pathergy reaction and a systemic cyclosporine intake.

Conflict of interest

The authors declare no potential conflicts of interest.

Acknowledgements

None.

References

1. McGarity WC, Robertson DB, McKeown PP, *et al.* Pyoderma gangrenosum at the parastomal site in patients with Crohn's disease. Archives of Surgery 1984; 119(10): 1186–1188. doi: 10.1001/archsurg.1984.01390220064014
2. Barbosa NS, Tolkachjov SN, el-Azhary RA, *et al.* Clinical features, causes, treatments, and outcomes of peristomal pyoderma gangrenosum (PPG) in 44 patients: The Mayo Clinic experience, 1996 through 2013. Journal of the American Academy of Dermatology 2016; 75: 931–939. doi: 10.1016/j.jaad.2016.05.044
3. Afifi L, Sanchez IM, Wallace MM, *et al.* Diagnosis and management of peristomal pyoderma gangrenosum: A systematic review. Journal of the American Academy of Dermatology 2018; 78: 1195–1204. doi: 10.1016/j.jaad.2017.12.049
4. Davis C, Wright B. Healing of a pyoderma gangrenosum at the site of a percutaneous endoscopically sited gastrostomy tube without tube removal. Case Reports 2014; 2014: bcr2014204301. doi: 10.1136/bcr-2014-204301
5. Lockenhofer F, Wollina U, Salva KA, *et al.* The PARAELSUS score: A novel diagnostic tool for pyoderma gangrenosum. British Journal of Dermatology 2019; 180: 615–620. doi: 10.1111/bjd.16401
6. Hashizume H, Kageyama R, Umayahara T, *et al.* A case of pyoderma arthritis, pyoderma gangrenosumu, and acne (PAPA) syndrome successfully treated with combination therapy of corticosteroids, cyclosporine, and colchicine. Trends in Immunotherapy 2018. doi: 10.24294/ti.v2.i2.719
7. Okuno A, Kanda Y, Nakano H, *et al.* Therapeutic effects of cyclosporine on Hailey-Hailey disease. Trends in Immunotherapy 2019; 3(1): 58–61. doi: 10.24294/ti.v3.i1.1140