Templates for Interesting Cases, Plastibytes and Review Articles

Interesting Cases:

Title

Authors and Affiliation

Keywords (3-5)

Case Description (limited to 50 words)

Questions (4 questions about essential aspects of the case)

Discussion (4 paragraphs, each one corresponding to the question asked)

Summary (one paragraph limited to 50 words)

References (limited to 8)

Below is an example Interesting Case Submission:

Interesting Case Series

Skin grafting in pyoderma gangrenosum

Marco Romanelli, M.D. Ph.D. Agata Janowska, M.D. Teresa Oranges, M.D. Valentina Dini, M.D., Ph.D.
Department of Dermatology, University of Pisa

Key Words: pyoderma gangrenosum, allograft, pathergy, adalimumab

Figure 1
Figure 2
Figure 3
Figure 4

CASE DESCRIPTION

A 75-year-old woman presented with multiple painful necrotic ulcers of the lower leg lasting since two months (Figure 1). Doppler studies of the venous and arterial system were within normal limits. The diagnosis of pyoderma gangrenosum (PG) was based on clinical and histopathological presentation showing a dermal aspecific neutrophilic infiltrate (Figure 2). Current blood screening with CBC, renal and liver function test, lipid profile, serum electrolytes, serum electrophoresis, clinical antibody profile, serology for rheumatoid factor, thyroid function test and blood sugar evaluation were within normal limits. The patient had a history of pathergy after antibiotic injection. Comorbidities were hypertension, atrial fibrillation and rheumatoid arthritis in remission. No history of allergies. A digital camera and a 3D measurement, imaging and documentation system (Star Aranz™) (Figure 3) were used to take the photos and to provide the precise measurement of size and healing trend evolution. The patient was followed up until the complete resolution of the disease. PG was managed with 0,5 mg/kg of

oral methilprednisolone, with local wound care, based on the principles of the wound bed preparation (1) and inelastic bandage therapy (2). For the first 3 weeks the local treatment was characterized by autolytic debridment using hydrogel (*NU-GEL*™), binding bacteria dressings (*Cutimed*® *Sorbact*®) 3 times per week to reduce the necrosis and fibrin. The wound care was continued with hydrofiber (Aquacel®) and inelastic bandage therapy twice per week for 2 weeks. Due to the worsening hypertension, the oral corticosteroids therapy was tapered, then interrupted. We did not use cyclosporine because of the hypertension and we performed anti TNFα therapy (adalimumab) 40 mg weekly (3). After one month using adalimumab, hydrofiber (Aquacel®) and inelastic bandage, the ulcers improved with more granulation tissue, with a reduction of the inflammation and the patient reported a reduction of pain level. The ulcers were characterized by superficial granulation tissue, absence of infection, critical colonisation or inflammation. We decided to perform an allograft due to pathergy history and to the wound bed clinical aspects. The allograft came from a cadaver skin and was cryopreserved at -80°C. The ulcers were cleaned with saline solution and antiseptic solution. The allograft was meshed 3:1 and fixed on the wounds with steri strip skin closure and then covered with binding bacteria dressing and inelastic bandage. We provided the procedure on the same day as a "day surgery" treatment. The follow up was performed twice a week for 3 weeks. We observed a perfect engraftment of the allograft during the final visit (Figure 4).

QUESTIONS

1. **What is pyoderma gangrenosum?**
2. **How is it diagnosed?**
3. **What is pathergy?**
4. **How is it treated?**

DISCUSSION

Pyoderma gangrenosum (PG) is a rare painful neutrophilic dermatosis that involves the skin and other organs. The diagnosis is based on the presence of suggestive clinical and histological aspects and the exclusion of other conditions. Typical histological evaluation shows epidermal ulceration, sterile neutrophilic infiltration without vasculitis or without granuloma formation but it can be often aspecific (4). Pathergy can occur in 30% of patients with PG. Pathergy is a condition in which a minor trauma can cause a development of the PG at the site of trauma (5). Considering this aspect is not recommended to perform surgical debridement in this condition and is mandatory to consider that surgical procedures in any anatomical site may induce the pathergy phenomenon. PG is extremely difficult to treat and it often needs the association of systemic treatments combined with a correct wound bed preparation approach (6). Corticosteroids are considered the first-line therapy for severe disease and cyclosporine is a good second-line option in patients with no hypertension or renal impairment. Other options include other immunosuppressive drugs,

intravenous immunoglobulin and targeted therapies, such as anti-TNFα, anti-interleukin 1, antiinterleukin 12,23 (5,7,8).

SUMMARY

A non invasive procedure like allograft was preferred due to the pathergy history of the patient. Other surgical approaches have also been described in pyoderma gangrenosum, such as the use of negative pressure wound therapy associated with
split thickness skin grafting under adequate immunosuppression (9). A combination therapy of anti TNFα and allograft can improve a graft engraftment with a reduction of immune response against the allograft. In this report allograft and anti TNFα (adalimumab) represented an effective association therapy for hardto- heal pyoderma gangrenosum.

REFERENCES

1) Schultz GS, Sibbald RG, Falanga V, Ayello EA, Dowsett C, Harding K, Romanelli M, Stacey MC, Teot L, Vanscheidt W.Wound bed preparation: a systematic approach to wound management.Wound Repair Regen. 2003 Mar;11 Suppl 1:S1-28. Review.

2)Konschawe W, Valesky E, Stage H, Junger M, Evidence of compression therapy. Hautarzt 2017 Aug; 68(8): 625-631.

3) Soto Vilches F, Vera-Kellet C. Pyoderma gangrenosum: Classic and emerging therapies. Med Clin (Barc). 2017 Sep 20;149(6):256-260.

4) Marzano AV, Borghi A, Wallach D, Cugno M. A Comprehensive Review of Neutrophilic Diseases. Clin Rev Allergy Immunol. 2018; 54(1): 114-130.

5) Alavi A, French LE, Davis MD, Brassard A, Kirsner RS. Pyoderma Gangrenosum: An Update on Pathophysiology, Diagnosis and Treatment. Am J Clin Dermatol. 2017 Jun;18(3):355-372.

6) Quist SR, Kraas L. Treatment options for pyoderma gangrenosum. J Dtsch Dermatol Ges. 2017 Jan;15(1):34-41.

7) Dini V, Romanelli M, Bertone M, Talarico S, Bombardieri S, Barachini P. Improvement of idiopathic pyoderma gangrenosum during treatment with antitumor
necrosis factor alfa monoclonal antibody. Int J Low Extrem Wounds. 2007 Jun;6(2):108-113.

8)Patel F, Fitzmaurice S, Duong C, He Y, Fergus J, Raychaudhuri SP, Garcia MS,

Maverakis E. Effective strategies for management of pyoderma gangrenosum a comprehensive review. Acta Derm Venereol. 2015 May; 95(5); 525-531.
9) Pichler M, Larcher L, Holzer M, Exler G, Thuile T, Gatscher B, Tappeiner L, Deluca J, Carriere C, Nguyen VA, Moosbrugger-Martinz V, Schmuth M, Klein GF, Eisendle K. Surgical treatment of pyoderma gangrenosum with negative pressure wound therapy and split thickness skin grafting under adequate immunosuppression is a valuable treatment option: Case series of 15 patients. J Am Acad Dermatol. 2016 Apr;74(4):760-765.