CASE REPORT

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Pemphigus foliaceus mimicking acute cutaneous lupus: A case report

Khalid Al Hawsawi[1]✉, Anas Tayeb[2], Suha Alsayed[3], Shahad Alkidaiwi[3], Abdulaziz Hussain Abdulmajeed[4], Abdulelah Hassan Alluhaybi[5]

[1]Department of dermatology, King Abdulaziz hospital, Makkah, Kingdom of Saudi Arabia
[2]Medical Intern, Graduate of the Faculty of Medicine, Umm Al-Qura University, Makkah, Saudi Arabia
[3]Medical Student, Faculty of Medicine, Umm Al-Qura University, Makkah, Kingdom of Saudi Arabia
[4]Medical Degree, King Fahad General Hospital, Jeddah, Saudi Arabia
[5]Department of dermatology, King Abdulaziz hospital, Makkah, Kingdom of Saudi Arabia

Authors' Contact Information

Anas Tayeb: anas-t@hotmail.com
Suha Alsayed: suhafakeeh@gmail.com
Shahad Alkidaiwi: shahad.alkidaiwi@hotmail.com
Abdulaziz Hussain Abdulmajeed: A.abdulmjeed@hotmail.com
Khalid A. Al-hawsawi: hawsawik2002@hotmail.com
Abdulelah Hassan Alluhaybi: Dr.abduleila@gmail.com

✉**Corresponding author**
Consultant dermatologist, Head of dermatology department, Department of dermatology, King Abdulaziz Hospital, Saudi Arabia
Email: hawsawik2002@hotmail.com

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General Note

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ABSTRACT

Pemphigus foliaceus (PF) is a rare form of pemphigus disease (PD). PD is a chronic autoimmune blistering skin disease. Here we report a case of a 65-year-old female who presented with a 2 months history of persistent slowly progressing painful skin lesions on her face. Skin examination revealed non-scaly, erythematous patches on her face with a butterfly rash distribution. Skin biopsy revealed subcorneal blister with presence of acantholytic keratinocytes in the blister cavity. Intracellular deposition of IgG and C3 in the epidermis was demonstrated using direct immune fluorescence (DIF). Complete blood counts, ESR, blood urea, creatinine, liver function tests, urine examination, ANA, ant d DNA, anti-smith antibodies were all within normal limits. Patient was diagnosed as Pemphigus foliaceus. We started her treatment using prednisolone 20 mg once per day. The lesions disappeared within few weeks of starting the treatment.

Keywords: Senear–Usher syndrome

1. INTRODUCTION

Pemphigus foliaceus (PF) is a rare form of pemphigus disease (PD). PD is a chronic autoimmune blistering skin disease in which auto antibodies are directed against the cell surface of keratinocytes. PD is divided into three major forms: pemphigus vulgaris, pemphigus foliaceus, and paraneoplastic pemphigus (Payne & Stanley, 2019). In PF, the auto antibodies are directed against desmogleins 1 causing loss of intraepidermal adhesions. It is commonly seen in middle aged patients with female predominance. PF appears clinically as scaly or crusted patches or plaques affecting sun exposed areas (the malar region of the face and in other "seborrheic" areas like scalp, upper chest and back. Small, flaccid bullae may rarely be present. Mucus membrane involvement is rare. Facial lesions have the typical butterfly rash distribution (Yang, 2014).

2. CASE REPORT

A 65-year-old female presented with a 2 months history of persistent slowly progressing painful skin lesions on her face. Past medical history was unremarkable. Review of systems did not reveal any history of oral lesions, photosensitivity, joint pain, or Reynaud's phenomena. There are no similar cases in the family. Skin examination revealed non-scaly, erythematous patches on her face with a butterfly rash distribution (figure 1). Hair, nail and mucous membrane examination was normal. Skin biopsy revealed subcorneal blister with presence of acantholytic keratinocytes in the blister cavity (figure 2). Intracellular deposition of IgG and C3 in the epidermis was demonstrated using direct immunofluorescence (DIF). Complete blood counts, ESR, blood urea, creatinine, liver function tests, urine examination, ANA, ant d DNA, anti-smith antibodies were all within normal limits. Patient was diagnosed as Pemphigus foliaceus. We started her treatment using prednisolone 20 mg once per day. The lesions disappeared within few weeks of starting the treatment.

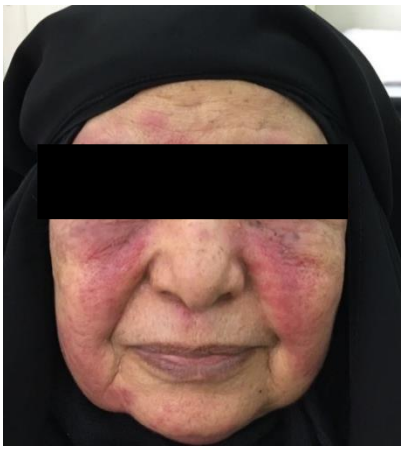

Figure 1 Diffuse, non-scaly, erythematous patches on the patient face.

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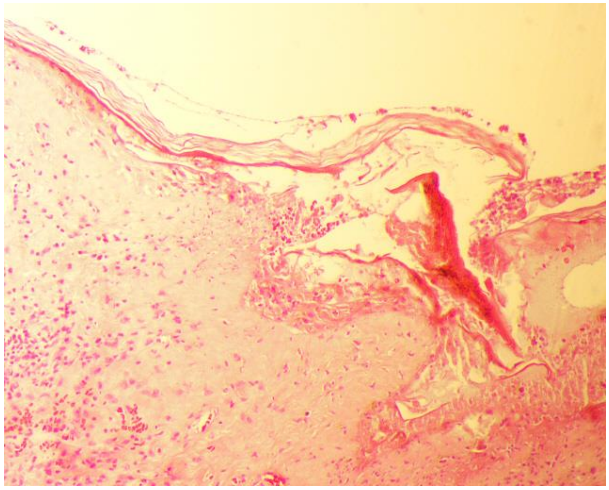

Figure 2 Skin biopsy showing subcorneal blister with presence of acantholytic keratinocytes in the blister cavity

3. DISCUSSION

Pemphigus foliaceus (PF) is a rare form of pemphigus disease (PD). PD is a chronic autoimmune blistering skin disease. PF presents clinically as scaly or crusted patches or a plaque affecting what is so called "seborrheic" areas that include the malar region of the face, scalp, upper chest and back. It is common in middle aged female (Försti et al., 2019). PF is common in middle aged women; however, our case is unusual considering the patient is 65 years old. Our patient showed atypical morphology of the rash in the sense that it didn't manifest as scaly or crusted erythematous patches, thus mimicking the presentation of acute cutaneous lupus. However, the butterfly rash distribution of the rash in our patient was typical for PF. The main differential diagnosis of rash in our patient includes acute cutaneous lupus, rosacea, PF and pemphigus erythematosus (PE). PE "Senear-Usher syndrome" is a variant of PF, has clinical features of PF but in addition to that, the patients have also circulating antinuclear antibodies of lupus erythematosus. The histopathology of PE shows features of both PF and cutaneous lupus. It shows subcorneal blister as well as interface dermatitis at dermo epidermal junction. The direct immunofluorescences (DIF) in PE have intercellular deposition of IgG and C3 both in the epidermis and in the basement membrane zone. Only few patients with PE have the two diseases clinically. Histopathology and DIF in our patient were typical for PF.

The main treatment of PF is systemic steroids. Other possible treatment options include dapsone, methotrexate, cyclophosphamide, and azathioprine. Avoiding the exposure of UV rays is crucial for the managing PE and PF (Igawa et al., 2004). Our patient responded well to small dose prednisolone..

4. CONCLUSION

Pemphigus foliaceus is an autoimmune disease, categorized as type of pemphigus disease. It resembles many diseases including PE, rosacea and acute cutaneous lupus. Careful examination and investigations are important as a rash in butterfly distribution is considered as one of the pitfalls in dermatology. The gold stander diagnostic tool for Pemphigus foliaceus is skin biopsy and DIF. Systematic corticosteroids are the mainstay of therapy.

Patient Consent

Informed consent was taken from the patient.

List of abbreviations used in the case report

Pemphigus foliaceus (PF), pemphigus disease (PD), direct immunofluorescence (DIF), erythrocyte sedimentation rate (ESR), Antinuclear antibody (ANA), deoxyribonucleic acid (DNA), Immunoglobulin G (IgG), lupus erythematosus (LE), Complement component 3 (C3)

Authors' Contributions

1. Dr. Khalid A. Al hawsawi, Dermatology consultant, MD (Principle Investigator and corresponding author) - Abstract, introduction, case report, discussion and conclusion.

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2. Anas Tayeb, MD (Co-Author) - Introduction and discussion
3. Suha Alsayed, MD (Co-Author) - Introduction and discussion
4. Shahad Alkidaiwi, MD (Co-Author) - Abstract and conclusion
5. Abdulelah Hassan Alluhaybi, MD (Co-Author) - Abstract and conclusion
6. Abdulaziz Hussain Abdulmajeed, MD (Co-Author) - Abstract and conclusion

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Conflicts of Interest

The authors have no conflicts of interest that are directly relevant to the content of this clinico-pathological case.

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