Pemphigus Foliaceus: Case Report and Review

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Introduction

Pemphigus foliaceus (PF) is a rare autoimmune blistering disease that can be difficult to diagnose and treat. The disease can be endemic or sporadic with a number of different instigating factors. We present a case of a 73-year-old male who developed PF shortly after treatment with 5-fluorouracil for actinic keratosis. The patient initially complained of itching and pain with 5-fluorouracil radiation treatment. The patient proved difficult to properly treat due to his transitory and resistant nature. A short review of PF is also provided.

Case Report

A 73-year-old male presented to our clinic for evaluation of persistent superficial erosions with overlying scale on his upper extremities, trunk, neck, and lateral face that began 3 months previous (Figure 1). The patient had just recently moved and was a new patient to us. At presentation, the lesions on the patient’s trunk and upper extremities had a thicker scale than those on his neck and face (Figures 2 and 3). Immediately prior to developing the lesions, the patient was treated with 5-fluorouracil for actinic keratosis on the face and scalp. The patient’s medication list included naproxen, clopidogrel, tamsulosin, and atorvastatin. Past medical history is significant for radiation therapy for laryngeal cancer and abdominal aortic aneurysm repair each many years previous.

Three lesional punch biopsies were performed for histology and eosin staining on chest and upper extremities. Histopathology showed separation of the cornel layer with subepidermal clefting of the granular layer, which favored the diagnosis of pemphigus foliaceus (PF) (Figures 4 and 5). Clinical correlation supported the diagnosis of PF. Direct immunofluorescence, indirect immunofluorescence, and ELISA were not performed due to logistical and clinical support of diagnosis, and particularly due to the patient’s resistance to further testing.

The patient was started on high-potency topical corticosteroids and a short course of 20mg prednisone daily. By the end of the first month of treatment, he was cleared of nearly all lesions. However, at the next follow up, the patient had a more localized flare up on the neck and lateral face while continuing the topical corticosteroids. The patient was resistant to starting any immunosuppressant medications other than corticosteroids. The patient was started on 1mg/kg per day of prednisone with the plan to taper. The patient is transitory and was soon lost to follow up after starting the new treatment plan.

Discussion

Pemphigus foliaceus (PF) is a member of the chronic blistering pemphigus family that includes other major forms: pemphigus vulgaris (PV) and paraneoplastic pemphigus. PF is a rare disease with low prevalence and incidence worldwide. PF is 2-3 times more common in women than in men. PF is a disease of the skin that presents as superficial erosions and crusting often located on the malar and superior facial region. The diagnosis of pemphigus foliaceus is based on histopathology demonstrating subepidermal vesicles. PF typically presents as red plaques on the head and neck, trunk, armpits, and hands. In early stages of the disease, the lesions may appear as superficial erosions and crusting. The lesions often involve the scalp and are characterized by superficial erosions and crusting.

PF can present with lesions anywhere on the body, but the most common sites are the face, neck, and upper chest. The lesions are typically asymptomatic or minimally symptomatic. The disease can affect people of all ages, but it is most common in adults over the age of 50. The disease can be acute or chronic, and in some cases, it can be recurrent.

PF is often associated with other autoimmune diseases, such as thyroiditis or rheumatoid arthritis. The disease is typically managed with immunosuppressive medications, such as corticosteroids, azathioprine, and mycophenolate mofetil. Although these medications can be effective, they can also have significant side effects, such as increased risk of infections and gastrointestinal bleeding.

The key to effective treatment of PF is early diagnosis and prompt initiation of therapy. Treatment options include topical and systemic corticosteroids, azathioprine, mycophenolate mofetil, and rituximab. The choice of therapy depends on the severity of the disease, the patient’s medical history, and the availability of local resources.

Conclusion

Pemphigus foliaceus is a rare disease that can be difficult to diagnose and treat. Our case highlights the complexity of PF and underscores the importance of prompt and accurate diagnosis. Early recognition and appropriate management are crucial for optimal outcomes.

References


Figure 1. Erythematosous plaques with erosions and crusting of the patient’s right face, ear, and neck.

Figure 2. White scaly plaques on patient’s bilateral dorsal forearms.

Figure 3. Scattered pink eroded plaques with some overlying crust on patient’s dorsal forearms.

Figure 4. I & E, original magnification 4x.

Figure 5. I & E, original magnification 10x.