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Exfoliative cheilitis related to psychologic factors uncovered during primary immunodeficiency evaluation

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Abstract

A 40-year-old previously healthy, non-atopic woman was referred for evaluation of a possible immunodeficiency disorder in the setting of an unusual erosive cheilitis and history of recurrent methicillin-resistant *Staphylococcus aureus* (MRSA) infection. Extensive work-up was non-diagnostic, including screening for immunologic disorders. She had failed multiple therapeutic modalities, including corticosteroid and immunosuppressive therapy. Tissue biopsy from the lip proved pivotal in demonstrating changes suggestive of factitial disease. This led to further detailed history-taking, yielding evidence of considerable psychologic distress. The patient was diagnosed with exfoliative cheilitis related to factitial disease in association with underlying untreated anxiety and psychologic trauma.

Keywords: anxiety disorder, exfoliative cheilitis, immunodeficiency

Introduction

Exfoliative cheilitis is a chronic superficial disorder of the lips that occurs most often in young women with a history of psychiatric disorders [1, 2]. A thorough medical evaluation is often necessary to exclude any primary or secondary immunodeficiency disorder.

Case Synopsis

A 40-year-old woman with a history of melanoma-in-situ and generalized anxiety disorder presented with a 2-year history of chronic swelling and painful peeling and crusting of the lips (**Figure 1**). In addition, she reported recurrent methicillin-resistant *Staphylococcus aureus* (MRSA) skin abscesses. She underwent several courses of topical, intra-lesional, and systemic corticosteroids, topical and systemic anti-fungals and antibiotics, and immunomodulatory and immunosuppressive therapies without improvement of her lips. Other than mild lymphopenia, the following laboratory workup was negative or within normal range: herpes simplex virus, varicella zoster virus, HIV immunoglobulin levels (IgE, IgA, IgM, IgG), adequate immune response to vaccine antigens, and negative genetic testing for common variable immunodeficiency. Desmoglein 1/3 and BP180/230 antibodies were negative, as was serum indirect immunofluorescence, thereby excluding autoimmune mediated bullous disease. Tissue culture from a lip biopsy was negative, although lip swab culture was positive for MRSA and *Candida*.

Recurrent MRSA skin abscesses and an unusual, treatment-resistant chronic cheilitis led to a presumptive diagnosis of a primary immunodeficiency disorder. Most notable on examination was the firm induration, worrisome for an infiltrative or unusual infectious process, which warranted an additional biopsy. This biopsy was



Figure 1. Physical examination findings consistent with an erosive and desquamative cheilitis. The figure on the left shows upper and lower lip enlargement with adherent crusting. The figure on the right highlights exudative crusting and exfoliative scale most prominent on the lower lip. There was marked induration and tenderness on palpation.

pivotal in the diagnostic process since the epidermal hyperplasia excluded diagnoses associated with a desquamating epidermis and the prominent subepithelial fibroplasia was suggestive of chronic changes as would be seen secondary to trauma (**Figure 2**). This diagnostic biopsy permitted a more focused conversation with the patient, which led to identifying a self-inflicted component to the lip peeling, a history of childhood sexual abuse, and a recent psychologically traumatic event in a family member that occurred in parallel with the exfoliative cheilitis. Following the diagnosis of exfoliative cheilitis with a factitial component in association with severe underlying psychological distress and a

known history of generalized anxiety disorder, the patient was referred for psychiatric consultation and ongoing psychological therapy was recommended.

Case Discussion

Exfoliative cheilitis occurs most commonly in young females, is believed to have a factitial component, and is typically associated with underlying psychological disorders [1,2]. The condition presents as lip swelling with persistent peeling and inflammation, manifesting as exfoliation, hemorrhagic crusting, and fissures [3]. The differential diagnosis of exfoliative cheilitis is broad, including inflammatory, immune-mediated, infectious, and neoplastic etiologies [1]. Also noteworthy is the fact of infection, which often suggests a possible immune dysfunction. However, recurrent infections in patients with exfoliative cheilitis might relate to a constellation of factors including skin barrier dysfunction and breakdown. Moreover, long-term use of immunosuppressive therapies, including corticosteroids as demonstrated in our patient, might increase the risk for secondary colonization and infection.

Treatment of exfoliative cheilitis is challenging and requires a full diagnostic evaluation to exclude other disorders and a comprehensive multi-disciplinary approach, including psychologic evaluation. Management of associated anxiety or depression is critical to successful management, as demonstrated in our patient. Therefore, exfoliative cheilitis is a

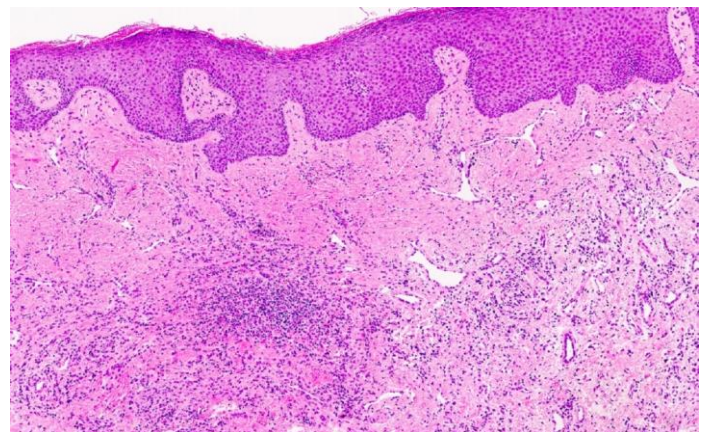


Figure 2. Punch biopsy, lip. Compact hyperkeratosis, hypergranulosis, epithelial hyperplasia and fibroplasia of connective tissue with mixed inflammation. H&E, 40x.

condition in which factitial, psychological, and secondary infectious etiologies are all relevant and must be considered, evaluated appropriately, and managed in a comprehensive fashion [4].

Conclusion

This case highlights the importance of obtaining an accurate and detailed history. The biopsy, which should exclude several conditions in the differential diagnosis may also show signs suggesting a factitial

etiology. Even in cases where an immunological diagnosis is entertained, it is important to perform a thorough psychiatric evaluation since patients in the dermatology and/or immunology consultation may often have a relevant psychological burden impacting their presentation.

Potential conflicts of interest

The authors declare no conflicts of interest.

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