Atypical Presentation of a Multisystem Disorder: Bullous Pemphigoid Masquerading as Pharyngitis

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Abstract

Bullous pemphigoid is a rare autoimmune blistering disease, presenting in older population with the classical skin blisters. We describe a case in which a 44 years old male presented without the typical skin manifestations of bullous pemphigoid making diagnosis extremely challenging. Accurate and timely diagnosis is the key to start appropriate treatment to prevent disabling complications of this disease.

Keywords: Bullous pemphigoid; Bullous presenting as pharyngitis; Bullous without skin blisters

Introduction

Bullous pemphigoid is not typically high on the differential diagnosis when a patient presents with presenting symptoms of odynophagia, pharyngitis, conjunctivitis, and oral mucosal ulcers. However with the development of new laboratory testing and a heightened clinical suspicion for the atypical presentation patterns, bullous pemphigoid is becoming more widely diagnosed in patients presenting with such atypical symptoms. The following case study aims to provide future directive to practitioners encountering nonspecific symptoms unexplained by commonly encountered pathologies, making the diagnosis of bullous pemphigoid less of a challenge.

Case Report

A 44 years old male with no significant past medical history presented in emergency room (ER) complaining of sore throat associated with odynophagia. His vital signs at presentation were: blood pressure (BP) 123/65 mm Hg, pulse: 78/min, temperature: 36.9 °C, respiratory rate 18/min, SPO2 98% on room air. Physical examination was unremarkable except for pharyngeal erythema. Lab work was significant only for ESR of 62. A diagnosis of pharyngitis was made and patient was discharged home on antibiotics. After 2 days at home, the patient presented to the ER with a white coated tongue and worsening sore throat. Examination was significant for oral thrush and persistent pharyngeal erythema. CT head and neck was done that showed soft tissue swelling in the pharynx. Patient was discharged home on antibiotics and fluconazole. Patient presented in the ER for the third time after 1 week, this time with redness of both eyes, dysphonia, along with worsening sore throat and odynophagia. Examination of oral cavity revealed multiple small oral ulcers with erythematous base. Eye examination showed redness with clear discharge and crusting of eyelids. Patient was admitted and started on ceftriaxone and acyclovir. Multiple tests were done to rule out all possible bacterial and viral causes showing negative results for rapid strep test, rapid plasma reagin (RPR), cytomegalovirus (CMV) IgM, Hepatitis panel, rheumatoid factor, ANA, HIV 4th generation, and Monospot. Throat culture showed no growth of beta-streptococci, blood culture likewise was negative for any growth. However both C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) were found to be significantly elevated. At this point, differential diagnosis was broadened to involve systemic disease as the cause of pharyngitis. Perilesional biopsy was done that showed histological diagnosis suggestive of bullous pemphigoid. And direct immunofluorescence showed linear immunoglobin IgG and linear C3 along the basement membrane suggestive of bullous pemphigoid. Patient was started on high dose steroids, topical and systemic. Over a couple of days, the patient showed significant clinical improvement. Sore throat and oral ulcers improved with clearing of sclera. Patient was discharged home on steroids.

Discussion

Bullous pemphigoid is an autoimmune blistering disease affecting older individuals. The classical findings in bullous pemphigoid are tense, fluid fill bullae on skin. It can affect any mucous membrane along with skin, mostly affecting oral cavity, ocular conjunctiva and external genitalia [1]. Very rarely it presents without classical skin blisters making diagnosis challenging as in our case report. Thus histology and immune florescence are mandatory to
make a definitive diagnosis and to rule out other diseases with similar presentation like other subepidermal dermatoses, such as linear IgA dermatosis, dermatitis herpetiformis, inflammatory epidermolysis bullosa acquisita and bullous systemic lupus erythematosus. DIF is the gold standard for diagnosis and demonstrates characteristic basement membrane zone antibody localization showing linear immunoglobulin G (IgG) and linear C3 staining in most of the cases [2]. Steroids are the mainstay of therapy but adjunctive agents are often added in an attempt to reduce long-term glucocorticoid dependence [3].

Conclusions

This case illustrates the potential for higher clinical suspicion of bullous pemphigoid with presenting symptoms of pharyngitis, conjunctivitis and oral ulcers. Recurrent pharyngitis or pharyngitis refractory to conservative measures should raise suspicion for a systemic disease as a manifestation of this symptom. Although these symptoms are not the typical presentations, they could represent a subset of patients presenting with atypical symptoms. Recognition of these atypical manifestations is crucial in rapid diagnosis and appropriate therapy to prevent complications such as neurological diseases and various malignancies associated with bullous pemphigoid [4, 5].

References