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Allergy and Immunology Second Opinion - Extended Written Report

Date: 2022-01-18

Patient: Jane Doe

Discussion:

61-year-old otherwise healthy woman presenting to dermatologist for pain and ulcers in the mouth and lips. Physical exam at that time revealing erythematous buccal mucosa and gingiva with many 1-1.5 cm ulcers. HSV PCR negative, CBC, CMP , ANA, ENA, RF negative. ESR 92 (high).

9/13/16 mucosal biopsy of left buccal area: suprabasal acantholysis with hemorrhagic cleft formation, basal cell hydropic degeneration and severe lymphohistiocytic infiltrate. No intranuclear inclusions (as in viral infections) identified. Histological findings are not specific but are consistent with paraneoplastic pemphigus

10/28/16 pathology review: differential diagnosis includes pemphigus vulgaris , oral manifestation of Darler's disease or possible oral manifestation of erythema multiforme. Intraepithelial vesicobullous disorder

Pt. was given prednisolone 20 mg and cortisone oral gel with good response to therapy and steroids were tapered off

The patient requests second opinion regarding treatment.

The patient has a new nonspecific diagnosis of a possible immune mediated dermatologic disease such as pemphigus vulgaris. The pathologic diagnosis, however, is not 100% certain at this time. Pemphigus is defined as a group of life-threatening blistering disorders characterized by acantholysis (loss of keratinocyte to keratinocyte adhesion) that results in the formation of intraepithelial blisters in mucous membranes and skin.

Patients with pemphigus develop mucosal erosions and/or flaccid bullae, erosions, or pustules on skin. This is a rare disease, usually occurring in adults with an average age of onset of 40 to 60 years. This is an autoimmune disease with acantholysis induced by the binding of autoantibodies to epithelial cell surface antigens leads to the clinical manifestations of pemphigus. The oral cavity is the most common site of mucosal lesions and often represents the initial site of disease. Most patients also develop cutaneous involvement manifesting as flaccid blisters on normal-appearing or erythematous skin. The Nikolsky sign (induction of blistering via mechanical pressure at the edge of a blister or on normal skin) often can be

elicited.

Treatment recommendations for this disease are to attempt systemic glucocorticoids initially along with oral glucocorticoids. Initial systemic steroids with prednisone are started at 1 to 2 mg/kg per day. In this case, the patient has experienced a complete response without evidence of disease and systemic steroids could be discontinued. If there is a relapse, systemic steroids should be started again in the same manner. If long-term steroid use is planned, other nonsteroidal systemic immunosuppressants such as azathioprine, MMF, and cyclophosphamide can be used to minimize the dose and length of treatment time with glucocorticoids. With systemic steroids studies have shown that 77% of patients responded to therapy.

More refractory cases can be treated with other immunomodulatory agents such as cyclophosphamide, rituximab, plasmapheresis, or intravenous immunoglobulins. This does not appear to be the case in this patient, but can be considered in the case of frequent and rapid relapse or lack of response to systemic glucocorticoids.

Recommendations:

1) Given complete response to therapy, continue observation at this time

2) If recurrence of disease, start prednisone again at 1 mg/kg daily with slow tapering after response. If long term steroids required, suggest addition of non steroidal immunosuppressant such as azathioprine or MMF

3) If refractory to the above, additional considerations may be cyclophosphamide, rituximab, IVIg, or plasmapheresis

4) Please reconsult as needed

Electronically Signed by: , MD on 01/18/2022 04:05:14 PM

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