JOURNAL OF DRUGS IN DERMATOLOGY
JDD
DRUGS • DEVICES • METHODS

Board Review

In partnership with Derm In-Review

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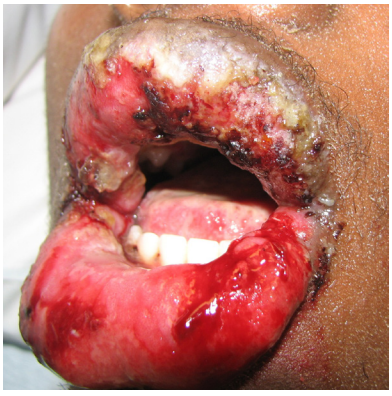
Jeremy Hugh, MD

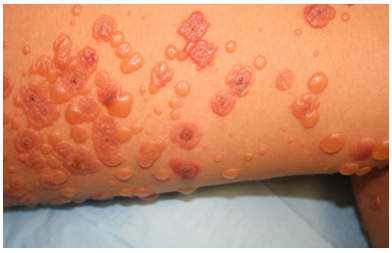
1)



A 16-year-old previously healthy male presents with severe mucositis as shown in the photograph, which developed over 1 week. He reported mild malaise and cough previous to this. He has no other cutaneous involvement (including vesicles, bullae, erosions, or targetoid lesions), but has conjunctival injection and crusting and sloughing in the nares. He denies recent unintentional weight loss, chills, or night sweats. His vital signs are unremarkable.There are no new medications and he has several negative HSV swabs and negative HSV antibody serologies. His CBC is unremarkable. Which of the following is the most appropriate next step in management, given the most likely diagnosis?

A. Start intravenous valacyclovir.
B. Order chest X-ray and mycoplasma serologies or PCR.
C. Transfer to burn unit.
D. Perform CT of head, chest, abdomen, and pelvis.
E. Start IVIG and aspirin.

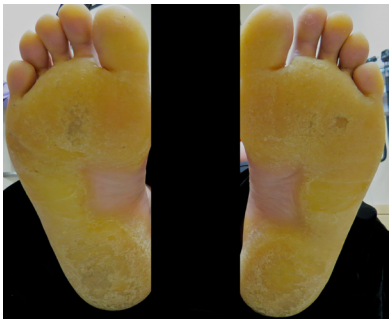
2)



A 17-year-old previously healthy female presents with blisters all over her body, including her oral mucosa, as depicted. She denies any new medications or recent infections. Which of the following is FALSE?

A. The most common antibody responsible for this binds to the NC16 domain of collagen XVII.
B. In drug-induced forms, vancomycin is more commonly a cause than penicillin.
C. Neutrophils are the predominant inflammatory component histologically.
D. The majority of these patients can obtain a clinical response to dapsone within 48-72 hours.
E. Eventual spontaneous resolution occurs in many patients.

3)



A 23-year-old female presents with asymptomatic thickened skin on her soles. She has mild thickening on her palms but denies any other skin findings. Upon further questioning, her mother, several maternal uncles, and maternal grandfather have had similar findings and some of them have also had esophageal cancer. Which of the following is FALSE?

A. The patient is also at risk for esophageal cancer, which typically occurs in the 5th decade.
B. This is inherited as an autosomal dominant disorder.
C. This patient likely has abnormalities in her hair as well.
D. This patient may also have oral leukokeratosis.
E. The cause of TOC is a mutation in the RHBDF2 gene.

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Board Review Answers

1) B. The correct answer is B. This is a case of Mycoplasma pneumoniae-induced rash and mucositis (MIRM).

Explanation/Literature Review

The main differential diagnoses include Stevens Johnson syndrome/toxic epidermal necrolysis (C), erythema multiforme (A), herpes simplex infection (A), paraneoplastic pemphigus (D), and less likely Kawasaki disease (E). Patients with MIRM typically have predominantly mucosal involvement (most commonly oral) and absent to sparse cutaneous involvement, which is often vesiculobullous or targetoid when present. It typically has a very good prognosis compared to SJS/TEN and is not caused by medications. CXR may show an atypical pneumonia and there is evidence of a Mycoplasma infection (IgM, PCR, etc.). It is useful to differentiate MIRM (aided by clinical findings and tests as above) from SJS/TEN to avoid unnecessarily aggressive treatment given the better prognosis. If workup for MIRM were negative, it would be prudent to evaluate for paraneoplastic pemphigus (D).

References: Canavan TN, Mathes EF, Frieden I, Shinkai K. Mycoplasma pneumoniae-induced rash and mucositis as a syndrome distinct from Stevens-Johnson syndrome and erythema multiforme: a systematic review. J Am Acad Dermatol. 2015 Feb;72(2):239-45.

2) A. The correct answer is A (A is false).

Explanation/Literature Review

This is a case of linear IgA bullous dermatosis, a subepidermal vesiculobullous disorder with direct immunofluorescence showing linear IgA deposits at the basement membrane. Clinically, tense bullae can appear annular, herpetiform, or in a linear fashion as a "string of pearls," as in this case. Antibodies are directed at a 97 kDa antigen near the collagenous domain of BPAg2 (BP180/Collagen XVII), in contrast to the MCW-1 region of the NC16 domain (near transmembrane portion) in other bullous disorders such as bullous pemphigoid (A).

3) C. The correct answer is C (C is false).

Explanation/Literature Review

Howel-Evans syndrome is an autosomal dominantly inherited disorder with focal palmoplantar keratoderma and squamous esophageal carcinoma. The PPK is typically worse on the soles and over areas of pressure. Abnormalities in hair and teeth have not been reported.

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