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DRUGS • DEVICES • METHODS

Senior Resident Peer-to-Peer Case Studies

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Case Studies authored by our educational partner GW School of Medicine and Health Sciences Dermatology Residency Program.

School of Medicine & Health Sciences
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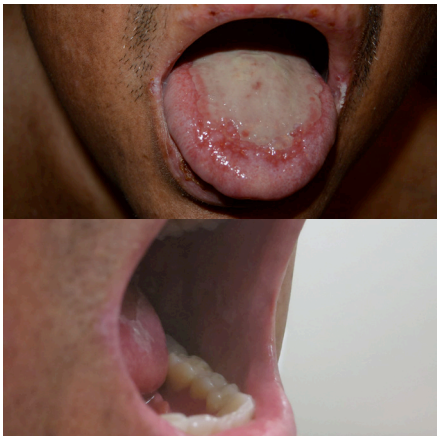

A 50-year-old Hispanic male presented with multiple erosions and ulcers on the mucosal lips and buccal mucosa. A broad ulcer was noted on the surface of the tongue (Images A and B). A biopsy was performed and demonstrated irregular acanthosis with saw-toothed rete ridges, a lichenoid infiltrate with predominately lymphocytes, baso-vacuolar degeneration of the dermal-epidermal junction, and cytoid bodies. Which of the following viral infections is most strongly associated with the patient's disease?

A. Epstein-Barr virus
B. Cytomegalovirus
C. Human papilloma virus
D. Hepatitis B virus
E. Hepatitis C virus

Explanation/Literature Review

The clinical and histologic findings are most consistent with erosive lichen planus. This variant of lichen planus (LP) affects the oral and genital mucosa with painful erosions and ulcers. The gingival can also be effected leading to gingivitis and periodontitis. When gingival disease is found together with involvement of the vagina and vulva, the term vulvovaginal-gingival disease has been used. The disease tends to affect middle aged to elderly adults and studies have shown that females are twice as likely to be affected when compared to males. Long term complications can include development of squamous cell carcinoma within the lesions and scarring of genital tissue leading the sexual dysfunction.

Of the variants of LP, the oral erosive type is most strongly associated with chronic hepatitis C virus (HCV) infection. A retrospective cross-sectional study from Brazil demonstrated that a statistically significant increase in the number of patients with oral erosive LP patient with HCV seropositivity compared to patients who were seronegative (30% versus 6.38%). Previous studies have also demonstrated the presence of HCV RNA when PCR tests were performed on tissue from oral lesions from 93.8% of seropositive patients. While the exact pathophysiologic process linking HCV infection and oral erosive LP is has not been completely elucidated, it is thought that the infection may produce to antigens that mimic proteins found on keratinocytes and give rise to an autoimmune reaction driven by memory and cytotoxic T-cells against the epidermis. Recently there have been multiple case reports documenting the successful use of direct acting anti-viral therapy against HCV with subsequent improvement or resolution of oral LP in seropositive patients.

The correct answer is E: Hepatitis C virus

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The content of these case studies, ideal to review during peer study groups, was developed by Julia Schwartz, MD and Thomas Lee, MD under the guidance of dermatologist Adam Friedman, MD, FAAD, Associate Professor of Dermatology, Residency Program Director, Director of Translational Research, Department of Dermatology GW University.

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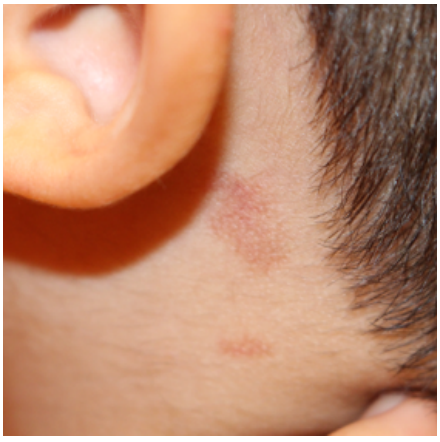

An otherwise healthy 1-year-old boy presented for evaluation of multiple, asymptomatic, red-brown, reticulated patches on the neck, torso and extremities that were present since birth (Images A and B). On further questioning, it was discovered that his older sister, father, paternal grandmother, and multiple paternal aunts, uncles, and cousins have similar birthmarks. Genetic testing revealed a heterogeneous deletion in RASA1. What is the most appropriate next step in management of this patient?

A. Pulsed dye laser
B. Magnetic resonance imaging of the brain and spine
C. Ultrasound at sites of skin lesions
D. Reassurance and observation
E. Radiographs of the skull

Explanation/Literature Review

The clinical photos show multiple capillary malformations (CMs) and based on the information provided in the question stem, a diagnosis of Capillary Malformation-Arteriovenous Malformation (CM-AVM) should be suspected. CM-AVM is an autosomal dominant syndrome characterized by the presence of multiple characteristic CMs with or without the presence of arteriovenous malformations (AVMs). The majority of families with CM-AVM have a heterozygous mutation in the RASA1 gene, which is a negative regulator of Ras, a crucial signal transducer for VEG-F mediated angiogenesis. Although the penetrance of this condition is > 95%, the number of CMs and the presence of AVMs is variable among affected family members.

The characteristic CMs are present at birth, multifocal, range from <1 to 3 cm, round or oval, and may be surrounded by a blanching halo. Rarely they can reach up to 15 cm and can be solitary. Fast-flow AVMs may be present in up to 30% and can occur in the brain, spine, bone, and muscle. In up to a third, the AVM involves the bone and soft tissue of the leg producing the phenotype of Parkes-Weber Syndrome. Intracranial and spinal AVMs occur in up to 7% and typically present by age 7 with symptoms ranging from seizures, hydrocephalus, headaches, cardiac failure and even death. Patients with suspected CM-AVM should be tested for the RASA1 mutation and have a screening MRI of the brain and spine to rule out the presence of AVMs that may be potentially life-threatening.

The correct answer is B: Magnetic resonance imaging of the brain and spine

References

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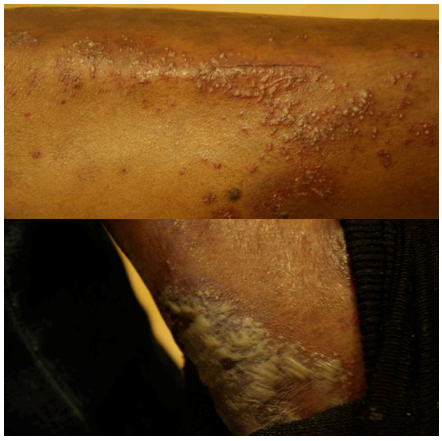

A 55-year-old female presented with an eruption of painful pustules on her bilateral upper and lower extremities and trunk. She notes that she has been suffering a scaly, erythematous rash for many years and three weeks ago she had received an injection of corticosteroids in the emergency room. On exam she had erythematous, scaly papules and plaques studded with coalescing pustules (Images A and B). A punch biopsy was performed and histologic examination showed areas of focal parakeratosis, hypogranulosis, acanthosis, and subcorneal neutrophilic aggregates forming spongiform pustules. No eosinophils were noted in the dermal inflammatory infiltrate. Bacterial and fungal stains were negative. Direct immunofluorescence testing was unremarkable. What complication could occur as a result of this skin eruption?

A. Hyperalbuminemia
B. Hypocalcemia
C. Hyperzincemia
D. Thyroiditis
E. Type 1 diabetes mellitus

Explanation/Literature Review

This patient's findings are consistent generalized pustular psorasis. This often occurs in the setting of acute withdrawal from systemic corticosteroids but can also occur from administration of certain medications such as beta blockers and lithium, which tend to exacerbate psoriasis. Studies show that only about 10% of patients have preceding history of psoriasis.

Electrolyte abnormalities can occur in generalized pustular psoriasis, in particular hypocalcemia, which, if severe enough, can lead to neurologic and cardiologic abnormalities. The mechanism is thought to be related to increased vascular permeability to serum proteins such as albumin, leading to hypoalbuminemia from loss to the interstitial space and urinary excretion. As around 40 percent of serum calcium is bound to these proteins, hypoalbuminemia results in hypocalcemia. Other cationic minerals such as zinc are also protein-bound, and as such patients may also develop hypozincemia. Consumption of zinc from rapid cell proliferation and turnover in psoriatic lesions may also contribute to the hypozincemia.

Autoimmune thyroiditis and type 1 diabetes mellitus can be sequelae of DRESS syndrome and are usually seen several months after resolution of the acute phase of the disease. These are not typically complications of pustular psoriasis.

The correct answer is B: Hypocalcemia

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