VIRGINIA DERMATOLOGY SOCIETY

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Chronic Mucocutaneous Candidiasis and the Relevance of EKG Monitoring

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A 24 year old female with a history of hypoparathyroidism presents for flare up of a chronic skin eruption since childhood. Physical exam revealed erythematous, scaly plaques on the arms, knees, and hands. The lesions began as small patches then progressed to larger plaques with fissures, bleeding and pruritis. She reports one twin sister with a similar condition and poor dental health.

Shave biopsy revealed hyperkeratosis with subcorneal neutrophils and subsequent PAS staining positive for hyphae. Based on the clinical findings, disease course, and family history, she was diagnosed with Autoimmune Polyendocrine Syndrome Type 1 (APS). She received oral fluconazole and was lost to follow up. Subsequently she returned to the clinic for further flare ups and had not taken fluconazole for 3 months. Serum chemistries revealed hypocalcemia with a level of 6.3mEq/L.

APS type 1 is caused by an autosomal recessive mutation in the autoimmune regulatory element (AIRE) gene which causes production of IL-17 antibodies and autoreactive T cells. As IL-17 is imperative for protection against candida, these patients have widespread mucocutaneous candidiasis[1]. Further workup includes genetic counseling, screening for associated autoimmune disorders (Addison's disease, hypoparathyroidism, diabetes, pernicious anemia) and oral examination for enamel hypoplasia. Female patients should receive fertility counseling as up to 60% of patients present with premature ovarian failure[2]. Dermatologists should obtain an EKG in all patients prior to administering oral fluconazole as hypocalcemia can contribute to prolonged QT Interval.

In this patient, once a stable EKG was obtained, we restarted fluconazole and she improved greatly.

1. Borchers J, Pukkala E, Mäkitie O, Laakso S. Patients With APECED Have Increased Early Mortality Due to Endocrine Causes, Malignancies and infections. J Clin Endocrinol Metab. 2020 Jun 1;105(6):e2207–13. doi: 10.1210/clinem/dgaa140. PMID: 32185376; PMCID: PMC7150614.
2. Husebye ES, Anderson MS, Kämpe O. Autoimmune Polyendocrine Syndromes. N Engl J Med. 2018 Mar 22;378(12):1132-1141. doi: 10.1056/NEJMra1713301. PMID: 29562162; PMCID: PMC6007870.