A Case of Polyglandular Autoimmune Syndrome Type One with Hypercalcemia and Hypotension

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Autoimmune polyendocrinopathy-candidiasis-ectodermal dystrophy (APECED) is also known as autoimmune polyendocrine syndrome type 1 (APS-1). We present a case of APS-1 with hypercalcemia of adrenal insufficiency during the calcium (Ca) treatment because of hypoparathyroidism.

Case: A 20-year-old female patient was diagnosed with APS-1 in 2004. She applied with the complaints of nausea and vomiting. Her laboratory findings proved hypercalcemia, hypernatremia, and hypotension. She had an upper respiratory infection a week prior to her admission to our hospital. Her echocardiography revealed increased pericardial brightness. Thus, indomethacin 2x1 was started as pericarditis was suspected. Blood pressure was 90/40 mmHg and the skin was dry. Horizontal ridges and trophic deformities on both finger nails and toes were observed. The laboratory findings were as follows: BUN 58 mg/dL, creatinine 0.82 mg/dL, sodium 127 meq/L, potassium 4.6 meq/L, Ca 12.6 mg/dL, follicle-stimulating hormone 4.42 mIU/mL, luteinizing hormone 5.63 mIU/mL, adrenocorticotropic hormone 124 pg/mL, cortizol 0.40 µg/dL, dehydroepiandrosterone sulfate 4.6 µg/dL, free triiodothyronine 1.94 pg/mL, free thyroxine 1.07 ng/dL, thyroid-stimulating hormone 3.26 µU/mL, anti-thyroid peroxidase 14.7 U/mL, parathyroid hormone 0.01 pg/mL. Hypercalcemia was associated with adrenal insufficiency. IV hydration and steroid of stress dose was started because of hypercalcemia and hypotension. We applied maintenance doses of hydrocortisone (30 mg a day) and fludrocortisone (0.1 mg x 1/2 a day) to the improved patient. Calcitriol and Ca were restarted to the patient as her Ca level was 6.5 mg/dL in the follow-up period. The patient had a leukoplakic lesion (candidiasis plaque) on the right buccal mucosa.

Discussion: APECED is a genetic disorder inherited in autosomal recessive tendency due to a defect in the autoimmune regulator (*AIRE*) gene located on chromosome 21.q223. In our case, hypercalcemia was thought to develop as a result of calcitriol/ and Ca treatment in addition to adrenal insufficiency. One should always remember that calcitriol and Ca treatment simultaneously with adrenal crisis triggered by stress assist the development of hypercalcemia in cases with APECED.

Key words: APECED, adrenal insufficiency, *AIRE*, hypercalcemia, hypoparathyroidism