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Sagliker Syndrome: A Rare Complication of Secondary Hyperparathyroidism

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Case Study: MC, a Hispanic female, presented to an ER with fatigue shortly before her 19th birthday. She was found to have a BUN of 234, creatinine of 17.5, hemoglobin of 1.8, calcium of 9.4, and PTH level of 3843. Imaging showed very small bilateral atrophic kidneys, extensive vascular calcifications and skull changes. Bone marrow biopsy revealed extensive fibrosis, active osteoblast/osteoclast activity, and absent iron stores. These were felt to be due to chronic metabolic change from secondary hyperparathyroidism (SHPT). She was very small in stature but reported normal growth until the age of 16. She was initiated on dialysis but due to her undocumented status, she was unable to receive scheduled dialysis, thus having dialysis only once or twice a week on an emergent basis. She was also started on cinacalcet for her SHPT. A parathyroidectomy was considered, but deferred pending improvement in other co-morbid conditions, including a large pericardial effusion. Over the next two years her PTH levels continued to be elevated, often over 5000. Her phosphorus levels remained elevated despite high sevelamer doses and calcium levels fluctuated, requiring decreases in the cinacalcet dose when hypocalcemic. She continued to have skull changes, especially in her maxilla, dentition, nose and mandible. During an episode of respiratory failure, intubation was difficult, requiring an emergency tracheostomy, which was permanent. After her tracheostomy she was no longer considered a candidate for parathyroidectomy and her skull changes worsened.

Problem: Sagliker Syndrome was first described in the literature in 2004 by Sagliker et al. He identified patients with uglifying human face appearance who had received insufficient treatment for SHPT in the earlier stages of Chronic Kidney Disease (CKD), often in developing countries. The patients were small in stature, had severe craniofacial deformities, very high PTH levels, hyperphosphatemia, hypocalcemia, and in many patients, depression. The incidence of Sagliker Syndrome is felt to be in only about 0.5% of the CKD population. Further research has found the possibility of a gene mutation.

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