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Leontiasis Ossea - A Case Report on Sagliker Syndrome in a 55-Year-Old Female

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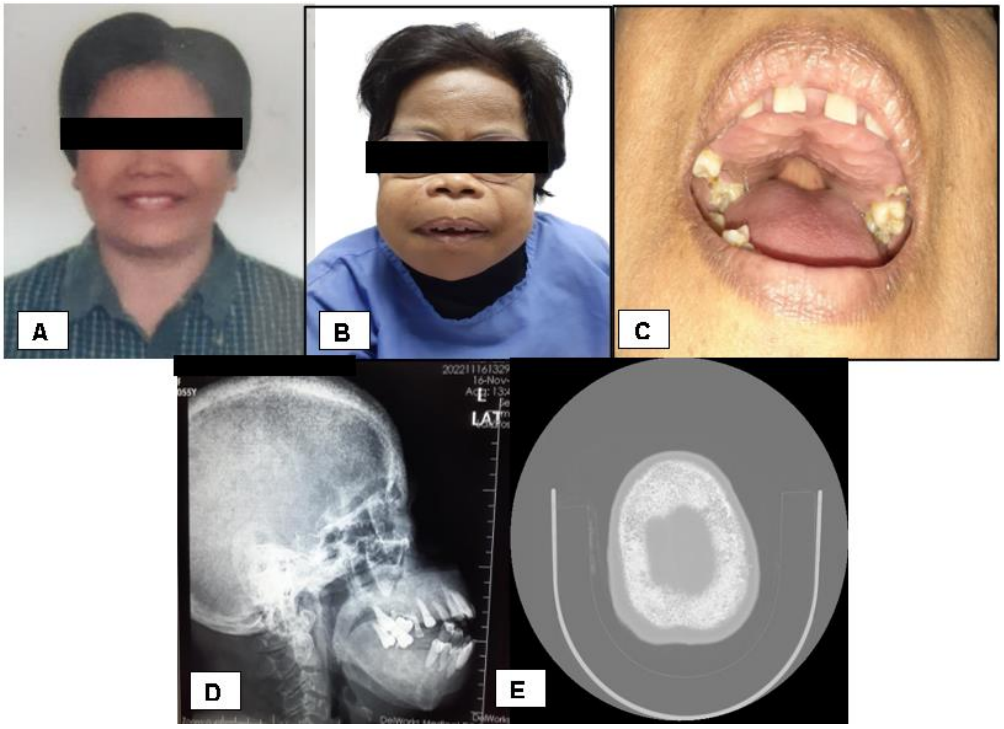
Case Study : Background: Sagliker Syndrome from untreated secondary hyperparathyroidism (SHPT) in end-stage renal disease rarely occurs. To our present knowledge, this is the first local case report of renal osteodystrophy with craniofacial deformities in a chronic kidney disease (CKD) patient on Maintenance Hemodialysis (MHD) in Eastern Visayas, Philippines. The Case: We report a case of a 55-year-old female, Filipino, with CKD Stage 5 on MHD from Chronic Glomerulonephritis. 8 months prior, she noted low back pain with difficulty in sitting and ambulating. Magnetic resonance imaging revealed multiple compression fractures with diffuse marrow edema at T12 to L3, probably secondary to osteoporosis but cannot rule out multiple myeloma or CKD Mineral Bone Disorder (CKD-MBD). 5 months prior, low back pain persisted, now with signs of SHPT in CKD or uremic leontiasis ossea (ULO) i.e., progressive jaw enlargement, teeth gap widening, and soft tissue accumulation of the soft palate. Patient expressed poor compliance with her maintenance medications. Pertinent laboratory workup revealed a significantly elevated intact Parathyroid Hormone (PTH) level at 3657pg/mL (15-65pg/mL) and phosphorus level of 1.81mmol/L (0.81-1.45mmol/L). Imaging studies revealed characteristic granular deossification or salt and pepper appearance of bones. Female sex, high phosphorus levels, and hemodialysis were notable risk factors in our case. A multi-disciplinary approach was advised, including hematology, otorhinolaryngology, and organ transplant surgery services. Diet, phosphate binders, reasonable use of vitamin D and calcium, adequate MHD with hemoperfusion, and parathyroidectomy were recommended to our patient. Complications of ULO like airway obstruction, difficulty in mastication, and poor quality of life were currently observed in our case. Conclusion: Our case highlighted that much is yet to be discovered about Sagliker Syndrome. Clinical and radiologic findings guided the assessment and management of the case. Risk factors should be identified early to prevent long-term complications since reversal is impossible after severe craniofacial deformities.

sagliker 1.png



sagliker 1.png

TEST	RESULTS	UNIT	NORMAL VALUES
Ionized Calcium	1.35 H	mmol/L	0.80 – 1.20
Phosphorus	1.81 H	mmol/L	0.81 – 1.45
Alkaline Phosphatase	584.6 H	U/L	0 – 110
PTH	3657 H	pg/mL	15 – 65