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ENDOCRINE IMAGING

Uremic leontiasis ossea

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A 36-year-old man with chronic kidney disease (CKD), undergoing hemodialysis therapy since 16-year-old, was referred to our hospital for the management of renal osteodystrophy. Fifteen years ago, he began to develop progressive painful changes in his body and face (Fig. 1a, b), especially massive enlargement of the jaws and palate (Fig. 1d, e). Substantial thoracic cage deformities resembling "pigeon chest" appearance (Fig. 1c), loss of height (171 to 160 cm), and shortening and clubbing of the distal phalanges were observed (Fig. 1f). Previous history also included chronic fistulized osteomyelitis of the left jaw, which appeared 5 years ago after a tooth extraction procedure, and fractures in the right clavicle and bilateral femur fractures, requiring the use of crutches to walk. Blood test results on admission showed total calcium level 9.2 mg/dL, phosphorus 8.3 mg/dL, 25-hydroxy-vitamin D of 27 ng/mL, alkaline phosphatase 3534 IU/L (normal range, 40–150 IU/L), and intact parathyroid hormone (iPTH) 3329 pg/mL (normal range, 10–65 pg/mL). Biochemical analysis obtained from the satellite hemodialysis unit of the last 5 years showed that the levels of serum calcium remained within the reference values, sometimes tending to mild hypocalcemia, but at any time it evolved with hypercalcemia. Besides, serum phosphate was mostly >5.5 mg/dL, alkaline phosphatase remained between 2000 and 3000, and iPTH was consistently >2500 pg/ml. The therapeutic approach with phosphorus binders and vitamin D analogues was late and ineffective, and the adherence of the patient was poor. Typical radiographic changes related to hyperparathyroidism (HPT) were identified in the patients' examinations, such as "rugger jersey spine", "salt and pepper" appearance in the skull, acro-osteolysis of the hands, brown tumor, and vascular calcification (Fig. 2). In addition, abnormal foci were visualized with 99mTc-sestamibi scintigraphy washout imaging, compatible with parathyroid glands. This case illustrates impressive bone deformities resulting from severe unremitting HPT in a dialysis patient, specially affecting maxillofacial bones, characterizing a rare manifestation of renal osteodystrophy known as uremic leontiasis ossea (ULO) [1, 2]. Secondary HPT is a frequent and complex metabolic disturbance encountered in the management of patients with CKD. The pathophysiology involves phosphorus retention, vitamin D deficiency, increased levels of fibroblast growth factor 23, and tendency to have low calcium levels, changes that collectively lead to a progressive hyperplasia of the parathyroid glands and increase in iPTH levels, in addition to vascular calcification. If not treated early, this condition can evolve with severe bone deformity and cardiovascular disease, and has high impact on the morbi-mortality of dialysis patients. Dietary phosphorus

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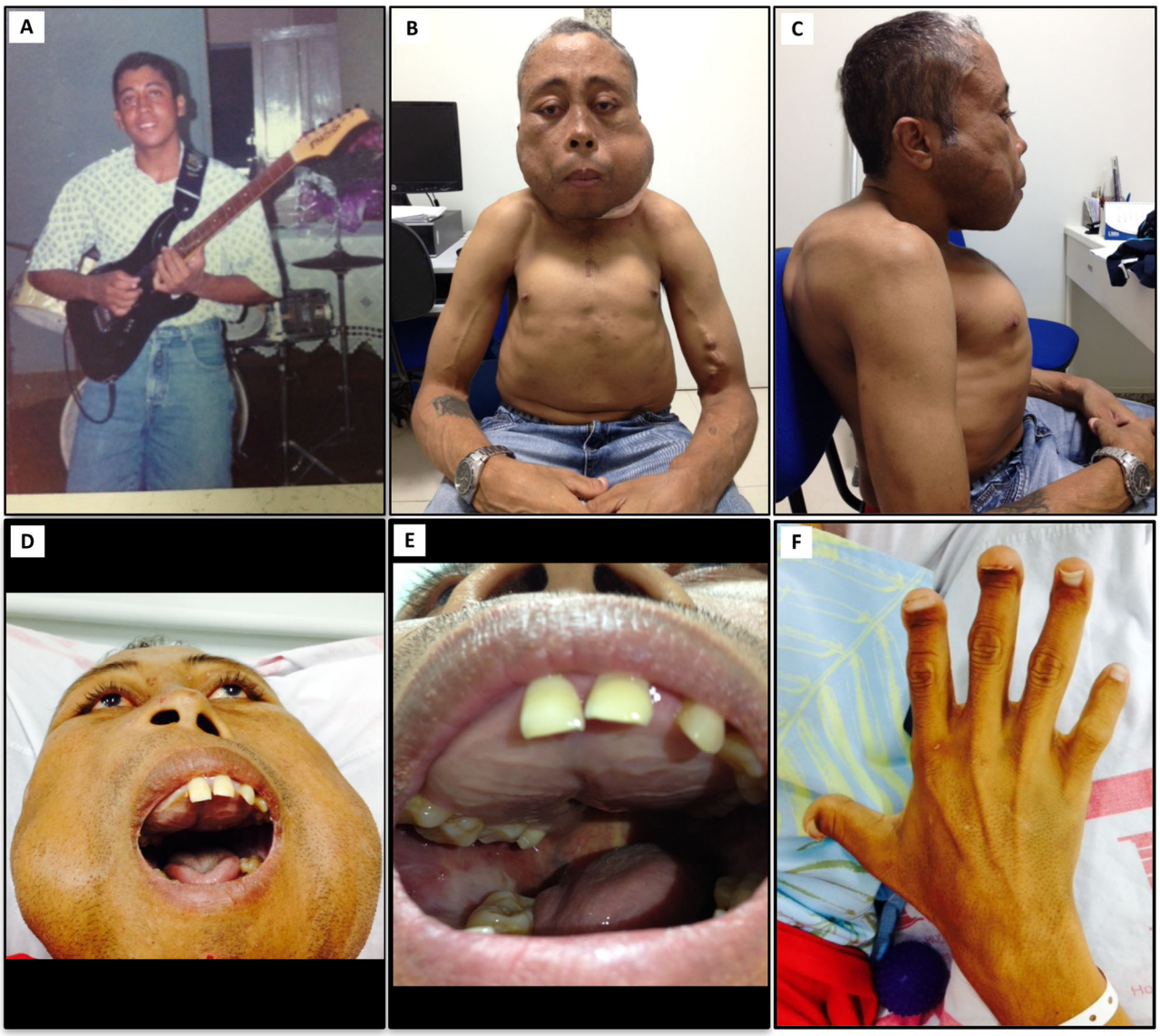

Fig. 1 Photos of the patient before **a** and after **b** the development of bone abnormalities. **c** Thoracic cage deformities resembling "pigeon chest" appearance. **d**, **e** show a marked enlargement of the palate and jaw bilaterally due to brown tumor. **f** The hands had shortening and clubbing of the distal phalanges

restriction and phosphate-binding agents, analogues of vitamin D and calcimimetics (such as cinacalcet) can be used in the treatmet, but in advanced cases parathyroidectomy may be necessary [3]. Given that normal craniofacial development often continues up until the 20s, it is conceivable that increased susceptibility to parathyroid overstimulation at a young age may be an important risk factor for the development of ULO in humans [2]. In this regard, we speculated that the evolution of our patient with ULO was linked to long-lasting uncontrolled HPT since young age along with suboptimal therapeutic management. Considering the occurrence of severe unremitting HPT and osteodystrophy, no drug therapy would be feasible for the patient and total parathyroidectomy with autotransplantation of parathyroid fragments into the forearm was performed. iPTH level was 17 pg/mL in the immediate postoperative period and 290 pg/mL after 1 year. Two of the four parathyroid glands are seen in Fig. 2f, and the largest of them had 6000 mg (standard weight is typically <50 mg). There was bone pain relief soon after surgery, but the osseous deformities regression was not relevant during the follow-up. Chronic osteomyelitis was partially controlled with long courses of antibiotic therapy and hyperbaric oxygen therapy sessions.

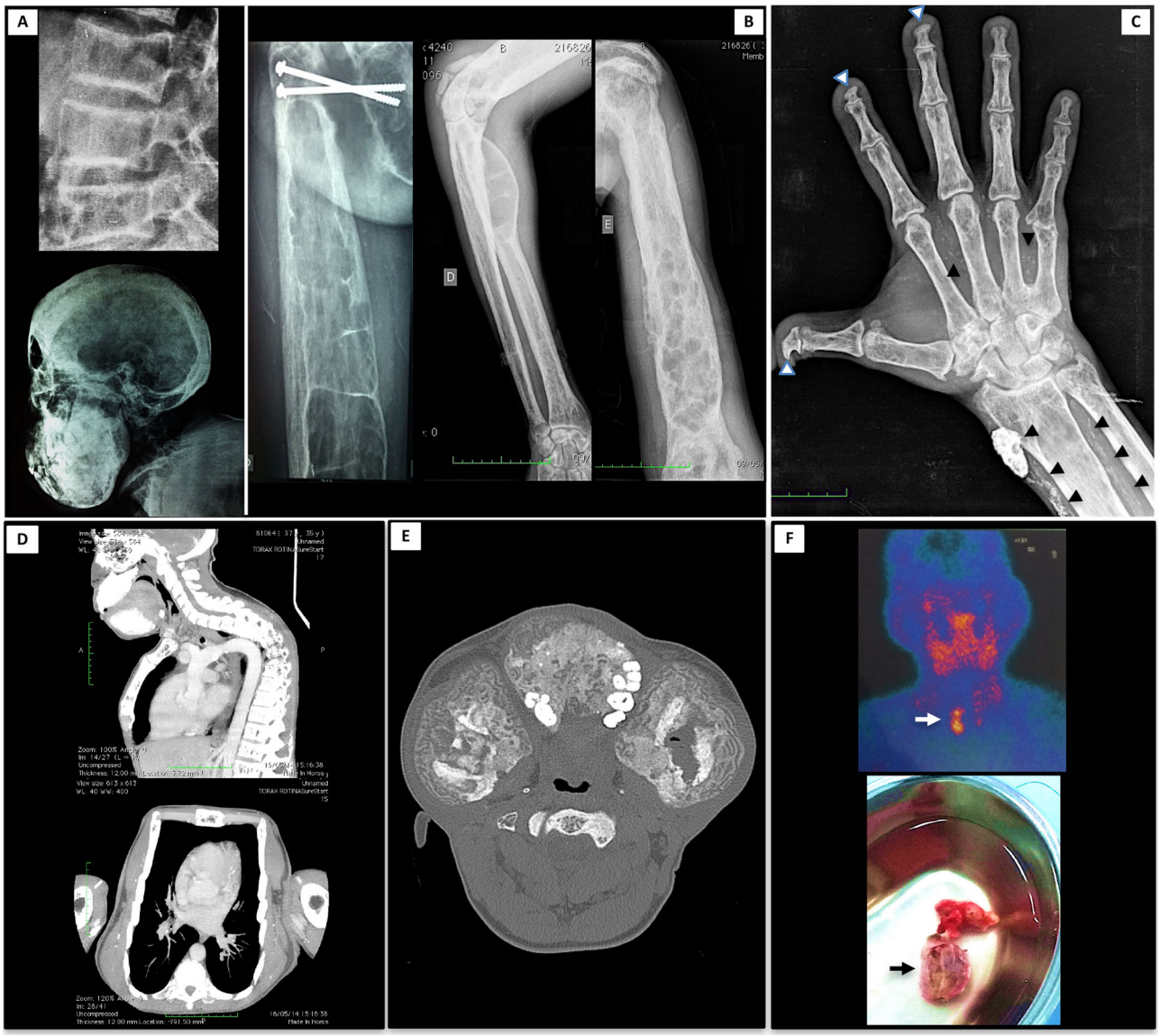

Fig. 2 **a** "Rugger jersey spine": sclerotic bands along the superior and inferior vertebral body with a relative band of lucency at the center. Skull X-ray showed the typical "salt and pepper" appearance, with thickening of the diploic space and multiple areas of osteolysis. **b** Radiographs of the femur (previously fractured), tibia, and humerus revealed multiple expansive osteolytic lesions, classically described as osteitis fibrosa cystica and brown tumor. **c** Radiographs of the hand showed multiple sites of subperiosteal resorption affecting the head of metacarpals and phalanges, resulting in severe tuftal resorption (acro-osteolysis, white arrowheads). The black arrowheads indicate vascular calcifications. **d**, **e** Computed tomography revealed deformities of the thoracic cage and lucent serpiginous tunneling within the maxillofacial bones with thinning of the cortical bone. **f** (above) White arrow: abnormal foci visualized with 99mTc-sestamibi washout imaging, compatible with parathyroid glands. **f** (below) shows two of the four parathyroid glands removed; the largest of them (black arrow) weighed about two hundred times more than a normal gland

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Informed consent Informed consent was obtained from the patient to publish this report.

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