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Neuroradiology/Head and Neck Imaging *Case Report*

Uremic Leontiasis Ossea

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ABSTRACT

While renal osteodystrophy is a common complication of chronic renal failure which is caused by secondary hyperparathyroidism, it is rare that the bony changes result in a severe progressive overgrowth of the bones of the face such that the patient is at risk for breathing and feeding difficulties. When this occurs, it is called uremic leontiasis ossea and patients who suffer from this rare, severe complication of renal osteodystrophy may go undiagnosed or be misdiagnosed resulting improper management due to its limited discussion in the literature. We report a case of a 42-year-old man with end-stage renal disease who was unable to receive dialysis consistently for many years who was found to have a large hard mass on the palate and palate ulcers.

Keywords: Uremic, Leontiasis, Computed tomography

INTRODUCTION

Leontiasis ossea is a historical term used to describe a rare craniofacial abnormality wherein progressive facial bony expansion results in an appearance similar to lion's face. This condition primarily involves the maxilla with progressive overgrowth of the bones resulting in encroachment of the sinuses, nasal cavities, and the orbital cavity. Benign fibro-osseous conditions like fibrous dysplasia are the most common causes for this condition. Milder form of the disease can be seen in conditions such as Paget's disease, acromegaly, and metabolic conditions like renal osteodystrophy. While renal osteodystrophy is a common complication of chronic renal failure which is caused by secondary hyperparathyroidism, it is rare that the bony changes result in a severe progressive overgrowth of the bones of the face such that the patient is at risk for breathing and feeding difficulties.[1] When this occurs, it is called uremic leontiasis ossea and has characteristic imaging findings. We report a case of a 42-year-old man with end-stage renal disease and development of leontiasis ossea with focus on the imaging findings.

CASE REPORT

A 42-year-old man with end-stage renal disease due to a combination of diabetes mellitus and cardiovascular disease presented to the emergency room with shortness of breath. Due to his lack of health-care insurance and ineligibility for funding to receive outpatient dialysis, he presented several times to the ED for intermittent dialysis where he was admitted and dialyzed if his symptoms met criteria for emergent dialysis. He survived this way for many years. On this presentation for emergent dialysis, he suffered a cardiac event requiring intubation for a cardiac cath. It was during extubation that the clinician noted marked the prominence of the palate and mucosal ulceration. Once stable from the procedure, further history from the patient was elicited. The patient noted that the bulky

hard mass on his palate had been present for several years and grew slowly overtime. He also noted progressive swelling and disfigurement of his face and noticed a hard mass along his mandible which had also grown overtime. The mass was not associated with pain and because he did not have insurance, he did not go to the doctor for further evaluation. He did not know what was causing the changes in his face. Oral maxillary surgery was consulted and they requested a computed tomography (CT) maxillary face [Figure 1] to be performed for further evaluation. It revealed severe enlargement of the maxilla and mandible with a coarse, marbled appearance of the bony matrix seen as alternating wavy bands of lucency and sclerosis. The bony expansion was most prominent at the maxilla and mandible with a bulky expanded hard palate accounting for the mass seen during extubation. These abnormal bony changes were also seen in other bones of the face, though less prominent, and at the skull base.

On review of the patient's chart and its documentation during multiple prior admissions for emergent dialysis, additional imaging and laboratory tests were analyzed. The patient had a long history of significantly elevated creatinine levels up to 17.5 mg/dL (normal 67–1.17 mg/dL), low glomerular filtration rate (GFR), and elevated blood-urea-nitrogen up to 125 mg/dL (normal 6–26 mg/dL) documented for over 7 years. Extremely elevated parathyroid hormone (PTH) was also noted, which had also increased over the years and was now >5000 pg/mL (normal 15–35 pg/mL), possibly greater as more definitive quantitative analysis was not routine beyond 5000 pg/mL. There was progressive, marked elevation of alkaline phosphatase reaching 740 units/L (normal 40–129 nits/L) and new hypocalcemia that was seen only in the past 7 months. Serum calcium was borderline decreased ranging from 8.1 to 8.3 mg/dl over the past few months (normal 8.5–10.1 mg/dl). There was also documentation supporting severe cardiovascular disease resulting in heart failure and diabetes mellitus.

A review of CT abdomen [Figure 2a] reveals an atrophic appearance of the kidneys containing multiple small cysts

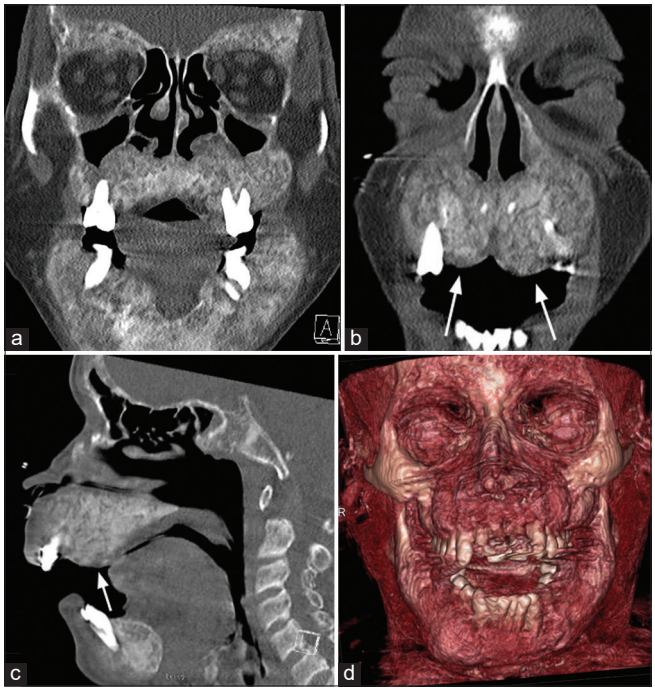

Figure 1: A 42-year-old man with uremic leontiasis ossea presenting with palatal mass. (a) Coronal CT image, bone reconstruction though the face showing marked expansion and marbled appearance of the bone, most prominently involving the maxilla and mandible but also seen involving the frontal bones around the orbit, (b) coronal CT image, bone reconstruction of the face more anteriorly shows marked expansion of the hard palate (white arrows). Note the board disfigurement of palate has the appearance of a lion's face, (c) sagittal CT, bone reconstruction showing prominence of the expanded hard palate visualized on physical exam, (d) 3D CT reconstruction of the bone showing the coarsened appearance of the expanded maxilla and mandible.

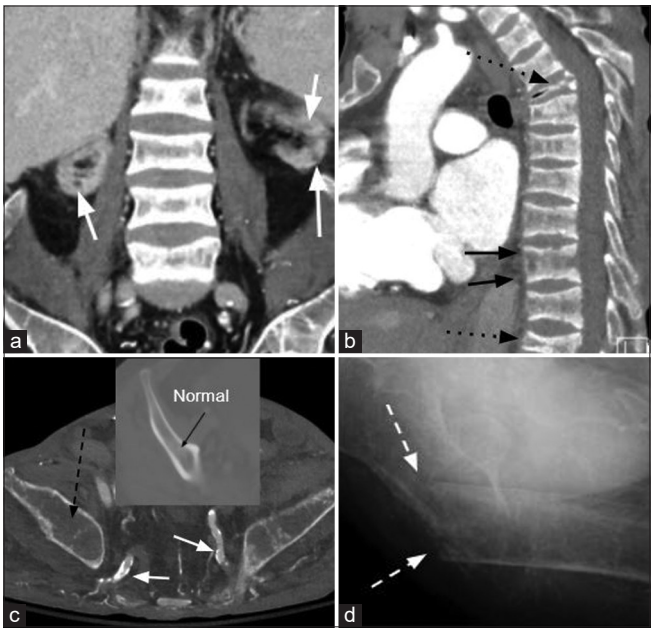

Figure 2: A 42-year-old man with renal osteodystrophy. (a) Coronal CT abdomen/pelvis with contrast shows bilateral atrophic kidneys containing multiple subcentimeter cysts (white arrows) characteristic of acquired cystic kidney disease in patients undergoing chronic hemodialysis, (b) sagittal CT chest with contrast showing multilevel bands of sclerosis along the endplates of the thoracic vertebrae (solid black arrows) with lucent bone in between, a characteristic appearance of metabolic bone disease in patients with hyperparathyroidism. Note vertebral compression fractures due lost bone mass (dotted black arrows), (c) axial CT of the pelvis showing dense calcification along the iliac arteries and marked osteopenia with loss of the normal medullary trabecular markings in the bones (black dotted arrow). The cutout image shows the appearance of normal trabecular markings (solid black arrow), (d) radiograph of the femur showing a femur fracture also found in our patient. There is angulation of the fracture fragments in the fracture bed. Fracture is a common complication of severe osteopenia.

typically seen in patients with end-stage renal disease who are on chronic hemodialysis and atherosclerotic calcification throughout his vasculature. Imaging [Figure 2b-d] also showed decreased bone density in the spine and pelvis and bands of sclerosis along the vertebral endplates, a characteristic feature of metabolic bone disease specific to hyperparathyroidism. In light of end-stage renal disease, laboratory and imaging findings as well as the history of progressive, painless expansion of the bones of the face over several years, uremic leontiasis ossea was diagnosed. After the cardiac event and subsequent cardiac cath, the patient left the hospital against medical advice.

DISCUSSION

Uremic leontiasis ossea is a rare, severe complication of end-stage renal disease and renal osteodystrophy. Renal osteodystrophy is a skeletal response to long-standing chronic renal disease and is caused by disruption in the balance of minerals and bone metabolism for which calcium, phosphorus, PTH, fibroblast growth factor 23 (FGF23), and Vitamin D are key players.[2] The health of the intestine, bone, and renal system is important in maintaining balance of these metabolites and hormones, and since most of the body's calcium and phosphate exists in bone as hydroxyapatite, the body can take steps to restore balance of the ions through hormone-induced bone turnover that, if persistent, will eventually change the strength and appearance of the osseous skeleton.[3]

Increased PTH concentrations first become evident when the GFR drops below 60 mL/min/1.73 m^2 and its increase is due to a decrease in the normal excretion of excess phosphate and a decrease in the presence of 25-OH Vitamin D, produced by the normal kidney.[1,2] Abnormally elevated PTH with the help of FGF23 will attempt to maintain normal serum phosphate levels by promoting continued excretion of excess phosphate from the body through the impaired kidney. It also maintains serum calcium balance, usually low due in chronic kidney disease, by releasing calcium from the bone. It does this by binding to PTH receptors on osteoblasts, which, in turn, increases osteoclast number and activity and results in bone resorption.[2] With progressive renal dysfunction, however, these compensatory mechanisms fail despite continued elevation of PTH.[3] Hyperphosphatemia then persists and worsens resulting in a further decrease in serum calcium. This hypocalcemia further stimulates the release of PTH to mobilize calcium, and with it, additional phosphate, from the bones at different sites in the skeleton.

Common bone changes in renal osteodystrophy caused by secondary hyperparathyroidism include osteitis fibrosa, osteomalacia, and osteosclerosis. Uremic leontiasis ossea is a severe form of osseous remodeling characterized by abnormal bone mineralization and severe, diffuse enlargement of the facial bones, particularly the maxilla and mandible, but also the skull base. Bone expansion results in facial disfigurement that appears in some cases similar to a lion's face. Leontiasis ossea is not unique to patients with renal disease and can be seen in a variety of disease processes known to result in enlargement of facial bones that give the broad appearance of a lion's face such as in Paget's disease, fibrous dysplasia, or gigantism.[4] As seen on imaging in our patient, the bones often appear marbled with alternating bands of hyperdensity and hypodensity throughout the bony matrix. This has been hypothesized to be due to necrosis and cystic degeneration representing the hypodense band and opaque osteosclerosis representing the hypersense band. This is similar to the well-known pattern multilevel band-like sclerosis seen along the vertebral body endplates of the spine with lucent bone sandwiched in between. When described, it is termed a rugger jersey spine given it close resemblance to the pattern seen on a rugby jersey. Brown tumors secondary to hyperparathyroidism can also occur in the mandible and though typically asymmetric and focal along the mandible as opposed to diffuse as seen in uremic leontiasis ossea, they too will often have this alternating pattern of lucency and sclerosis which is indicative of rapid osteoclastic bone turnover and repair thought to result in this marbled appearance.[5,6]

The greatest risk to patients with uremic leontiasis ossea is specifically related to the marked expansion of the maxilla and mandible and its ability to compromise the oropharyngeal airway and impair oral intake. Another significant risk to these patients is related to its potential for skull base involvement. Progressive bone expansion that gradually encroaches and narrows foramen or osseous canals through which important neurovascular structures travel can result in neurologic compromise that increases morbidity and mortality. For example, expansion of the bones surrounding the optic canal may compress the optic nerve and result in vision loss. Similarly, narrowing of the foramen ovale at the skull base may compress the traversing V3 portion of the trigeminal nerve resulting in sensory denervation of the skin and oral mucous membranes and/or motor denervation the muscles of mastication further impairing oral intake. Involvement of the carotid canal at the skull base if severe enough could result in progressive compression of the passing internal carotid artery and potentially increase the risk of stroke. Without knowledge of the potential adverse contribution of bony overgrowth at the skull base to severe neurologic complication in these patients, necessary discussion of the risks, treatment options, and prevention methods to mitigate such disaster cannot be performed.[4-6]

CONCLUSION

Uremic leontiasis ossea is a rare, severe complication of renal osteodystrophy that results in mark enlargement of the

facial bones and is infrequently described in the literature. Its appearance on imaging is similar to more recognized osseous changes of secondary hyperthyroidism including brown tumors and rugger jersey spine indicative a similar pattern of rapid bone turnover and repair, but the marked, diffuse involvement of the facial bones and cranial vault poses a unique set of risks to these patients, both known and likely unknown at this time. Increased awareness is paramount and will allow for further knowledge of its typical appearance on imaging and presentation as well as the unique risks/complications specific to these patients which are important for increased discussion of proper management.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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