Interhospital Endocrine Conference

Case 4

Uremic leontiasis ossea

Sagliker syndrome

Leontiasis Ossea

Lion face syndrome

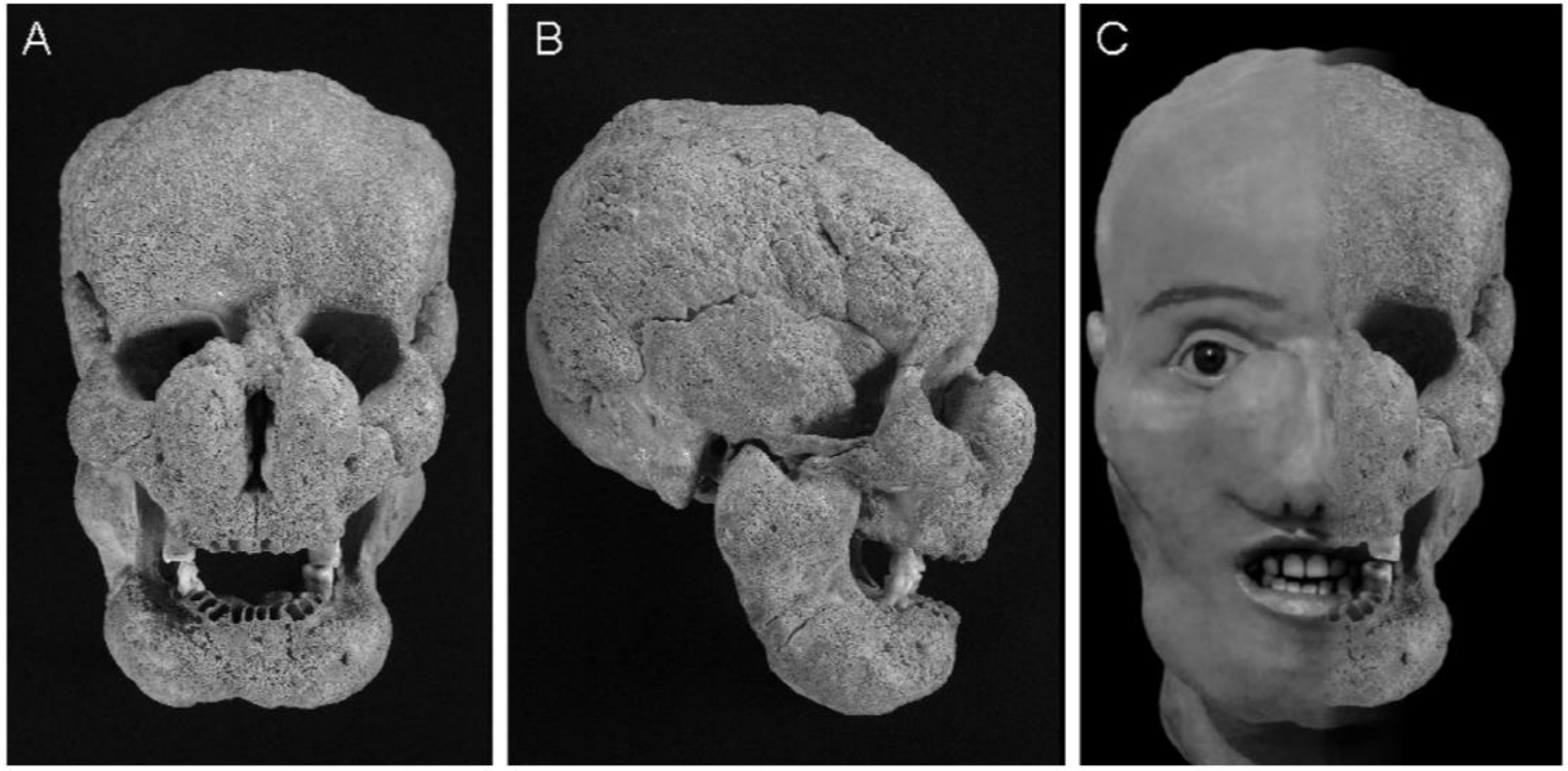
History

The medical term ***Leontiasis ossea*** was first used by ancient Greek physician Rufus of Ephesus to describe bone changes caused by disease that made the sufferer appear to have a **lion-like face.**

Kristina Killgrove Former Contributor *Archaeologist, Writer, Scientist,* Dec 27, 2017

Leontiasis Ossea

- Leontiasis ossea is a term used to describe the lion facies associated with excessive bone formation of single or numerous craniofacial bone.



Braz. J. Nephrol. (J. Bras. Nefrol.) 2019;41(2):304-305

Causes of Leontiasis Ossea

- Hyperparathyroidism and renal osteodystrophy
- Paget's disease
- Fibrous dysplasia (including McCune-Albright's syndrome)
- Craniometaphyseal dysplasia
- Metastasic disease
- Multiple osteomas (including Gardner syndrome)
- Osteogenesis imperfecta
- Acromegaly
- Neurofibromatoses
- Treponematosis (including syphilis, yaws, Goundou)
- Pycnodysostosis
- Hyperostosis frontalis interna (including Morgagni-Stewart-Morel's syndrome)
- Camurati-Engelmann's disease
- Endosteal hyperostosis (including Van Buchem's and Worth's types)

Braz. J. Nephrol. (J. Bras. Nefrol.) 2019;41(2):304-305

Leontiasis Ossea

- End-stage renal failure especially concurrent with continuous hemodialysis is a common cause of bone metabolism alteration
- Clinical condition face phosphate retention and reduced vitamin D conversion leading to hypocalcemia that induce PTH over-secretion
- Inadequate hemodialysis, failure to manage phosphate and calcium can result in severe hyperparathyroidism leading to marked osteodystrophy
- This condition is characterized by hyperostotic changes in the facial bones that can result in bilateral expansion of the malar processes
- Leontiasis Ossea in patient with severe hyperparathyroidism in end-stage renal disease patients, which has been less commonly reported due to dialysis and medical treatment advances in the last decade

Smile magazine, Maxillo-facial surgery, Lion Face Syndrome A Rare Case Report
Braz. J. Nephrol. (J. Bras. Nefrol.) 2019;41(2):304-305

Renal osteodystrophy

Facial skeletal changes associated with hyperparathyroidism in chronic renal disease are infrequently encountered and assume 3 radiographic patterns

1) The classic form is termed 'osteitis fibrosa cystica' and presents with a combination of increased osteoblast activity, peri-trabecular fibrosis, and cystic brown tumors.

2) The second form resembles fibrous dysplasia, exhibiting a classic ground-glass pattern on both conventional X-rays and CT scans.

3) The third and rarest form is known as uremic leontiasis ossea (Sagliker syndrome).

P. Dehghan et al. ‚Journal of Biosciences and Medicines, 2015, 3, 54-59

Uremic leontiasis ossea/ Sagliker syndrome

- Uremic leontiasis ossea is characterized by significant hypertrophy of the jaws, serpiginous 'tunneling' or channeling within the bone, and poor visualization of the cortical bone.
- Secondary hyperparathyroidism due to chronic renal failure and hemodialysis may cause a broad range of osteodystrophy changes, especially in the skull and facial bones, leading to a rare phenomenon known as uremic leontiasis ossea or bighead disease.
- Uremic leontiasis ossea may be caused by mutations in the GNAS1 gene, FGF23, or FGFR3.

P. Dehghan et al. ‚Journal of Biosciences and Medicines, 2015, 3, 54-59

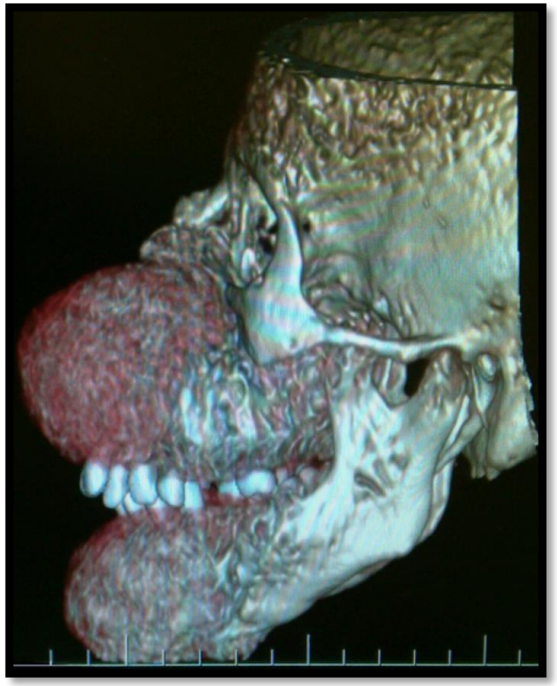
Lateral skull radiography

- Enlargement of both the maxilla and mandible, along with a ground-glass appearance and loss of corticomedullary differentiation, often occurs concomitantly with dental malocclusion and the absence of Lamina Dura.
- The oropharyngeal space exhibits narrowing.

Axial CT of the face

- Extensive bone growth with demineralization of the mandible, characterized by diffuse bone tunnels.

P. Dehghan et al. ‚Journal of Biosciences and Medicines, 2015, 3, 54-59



3D reformatted CT image

- Deformities of the maxilla and mandible, resulting in encroachment upon the orbital, oral, and nasal cavities.

Coronal CT scan of face and skull

- Hyperostosis of facial and cranial bones, along with a ground-glass expansion of the maxilla, mandible, and skull featuring bone tunnels.
- Marked hypertrophy of the hard palate resulting in significant narrowing of the oropharyngeal space.

P. Dehghan et al. ‚Journal of Biosciences and Medicines, 2015, 3, 54-59

Uremic leontiasis ossea/ Sagliker syndrome

- Combining calcium and vitamin D supplements with reduced phosphate levels has been recommended as effective.
- Parathyroidectomy stops further facial deterioration, although it may be complicated by hungry bone syndrome.
- Surgical correction of facial bones is controversial, and facial deterioration may stabilize or even improve mildly after parathyroidectomy, this underscores the importance of early recognition for prompt management and the prevention of severe disfigurement

P. Dehghan et al. ,Journal of Biosciences and Medicines, 2015, 3, 54-59
Braz. J. Nephrol. (J. Bras. Nefrol.) 2019;41(2):304-305

Case report

Sagliker Syndrome: Uglifying Human Face Appearance in Late and Severe Secondary Hyperparathyroidism in Chronic Renal Failure

Yahya Sagliker,[*,†] Mustafa Balal,[*] Piril Sagliker Ozkaynak,[†] Saime Paydas,[*] Cemal Sagliker,[†] Hasan Sabit Sagliker,[†] Necati Kiralp,[‡] Siddik Mumin Adam,[‡] Ilhan Tuncer,[*] Gulfiliz Gonlusen,[*] Mustafa Esenturk,[§] Erdal Gocmez,[§] Hulya Taskapan,[‖] Mehdi Yeksan,[¶] Ersin Kobaner,[#] Ozan Ozkaya,[**] Musa Yuksekgonul,[‡] Idris Emir,[††] Nurdan Cengiz,[‡‡] Ismet Onder Isik,[§§] Omer Bilginer,[§§] Turgay Guler,[‖‖] Hasan Yakar,[‖‖] Nedim Sarsmaz,[‡] Serkan Dilaver,[‡] Balim Akoglu,[‡] Mustafa Basgumus,[¶¶] and Ercin Chirik[##]

Almost every patient with chronic renal failure (CRF) eventually develops secondary hyperparathyroidism. Found **25 patients** who had CRF, SH, short stature, extremely severe skull changes, maxillary and mandibular bone changes, teeth/dental abnormalities, and soft and innocuous tumoral tissues in the mouth (hence, uglifying the appearance of the face), fingertip changes, severe psychologic problems, and depression.

Turkey, 2003

Case report

• A 25 year old Palestinian patient attended the author's clinic complaining of a huge bony growth in the facial bones, orbits, nose, mandible and skull.

Palestine

Lion Face Syndrome A Rare Case Report, Maxillo-Facial Surgery 2008

• A 36-year-old female patient who had been treated with chronic hemodialysis and who developed secondary hyperparathyroidism.

Serbia

HIPPOKRATIA 2015, 19, 3: 266-267

• A 59-year-old woman presented with a progressive facial deformity since several months ago. She had end-stage renal disease undergoing regular hemodialysis for 6 years.

Taiwan

QJM: An International Journal of Medicine, 2017, Vol. 110, No. 2

Case report

• A 62-year-old male patient diagnosed with chronic glomerulonephritis was maintained on hemodialysis for the previous 12 years

Nanjing, China

JBMR PlusVolume 2, Issue 4Jul 2018PagesF i-F iv, 187-245

• A 47-year-old female with CRF on hemodialysis for 6 years is referred from the endocrinology service to Maxillofacial Surgery Department with a chief complaint of severe maxillary and mandibular enlargement.

Chili

Journal of Stomatology, Oral and Maxillofacial Surgery, February 2018, Pages 56-60

• A 24-year-old woman presented with asymmetric facial deformity and stature shortening. She was diagnosed with end-stage CRF and had undergone regular peritoneal dialysis.

Shanghai, China

World J Clin Cases 2019; 7(22): 3792-3799

Case report

• A 39-year-old female with end-stage renal disease, undergoing hemodialysis for 11 years, was transferred from Angola to Portugal.
Angola, Southwest africa

Braz. J. Nephrol. (J. Bras. Nefrol.) 2019;41(2):304-305

• A 42-year-old man with end-stage renal disease who was unable to receive dialysis consistently for many years .
USA

Journal of Clinical Imaging Science • 2021 • 11(27)