Uremic Leontiasis Ossea: A Rare Presentation of Chronic Renal Failure Secondary to Hyperparathyroidism

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ABSTRACT
Facial skeletal changes associated with hyperparathyroidism assume three radiographic patterns: osteitis fibrosa cystica, fibrous dysplasia, and leontiasis ossea. The 3rd pattern is unique to renal osteodystrophy. Renal osteodystrophy frequently affects the spine, ribs, long bones, and skull. Findings of renal osteodystrophy in facial and cranial bones are rare. However, it’s most severe osseous complication is characterized by massive thickening of the cranial vault and facial bones, called uremic leontiasis ossea (ULO), with only few cases reported in the literature.
The uremic leontiasis ossea causes significant aesthetic and functional changes. It is important to recognize features of leontiasis ossea, as it may result in life threatening upper airway obstruction and compressive cranial neuropathy while after parathyroidectomy, facial changes can be stabilized or improved mildly.
We report a case of uremic leontiasis ossea with a history of gradual enlargement of the facial bones over a period of one year. Significant hypertrophy of the maxilla and clavarial bone is most significant CT finding with serpiginous tunneling within the bone and poor visualization of the cortical bone. Nuclear medicine scans are also useful for demonstrating parathyroid adenoma. Ultimately, the diagnosis of uremic leontiasis ossea can be made non-invasively through a combination of clinical parameters and imaging findings, as described in this article.

Keywords: Renal osteodystrophy, secondary Hyperparathyroidism, uremic Leontiasis ossea

1. INTRODUCTION
Leontiasis ossea also found in the literature as leontiasis or lion face, is a rare medical condition with characteristic overgrowth of the facial and cranial bones. The term leontiasis ossea is reported to be first coined in 1864 by the famed Virchow [1]. Leontiasis ossea is not itself a disease, but a condition of other diseases such as Paget’s disease, gigantism, fibrous dysplasia, hyperparathyroidism and renal osteodystrophy [1]. Diagnosis of the specific osteodystrophy type is a rather complex process and various biochemical markers and radiographic findings are used so as to facilitate this stage [2].
Findings of RO caused by secondary hyperparathyroidism (SH) in cranial bones are frequent and include osteomalacia, osteosclerosis, and erosion of the cortical bone, brown tumors, and resorption of the lamina dura [3]. During the last decade this uremic complication is less frequently reported in the literature. This is probably due to better dialysis and better medical control of the secondary hyperparathyroidism.

We report a case of 44-year-old male patient with a complex medical condition of end-stage chronic renal failure and secondary hyperparathyroidism presenting with a history of gradual enlargement of the facial bones over a period of one year.

2. CASE PRESENTATION

A 44 years old male presented to maxillofacial surgery outpatient clinic complaining of worsening facial deformity over the past year, now causing dental malocclusion and dysarthria. There was no reported pain, visual or neurological deficiencies associated with the enlarged facial bones. He had been on maintenance hemodialysis for 2 years when he became noncompliant with oral therapy. Clinical examination showed that the patient is of short stature and was confined to a wheelchair due to old pelvic bone and left humeral head fractures with bone pain related to the severe osteoporosis status. There was a significant marked non-tender maxillary and mandibular hypertrophy with nasal flattening and severe palatal changes, also loss of his front teeth along with spacing of upper and lower teeth. (Fig 1, 2).

The craniofacial computer tomography (CT) showed extensive calvarial and maxillofacial bones involvement specially hard plate and mandibles it characterized diffuse bone thickening with alternated pattern of osteolysis and osteosclerosis and low-attenuation serpentine “tunneling” extending through the bone that resembling a tabby appearance. The affected areas lacked clearly defined cortical bones with no corticomedullary distinction. (Fig 3, 4).

Figure 4: axial CT images shows sever thickening with bone tunneling involving the hard palate and mandible. There is virtually no cortical bone identified in the palate

A Panorex film showed decreased cortical bone and loss of the lamina dura around the teeth. Parathyroid scintigraphy was requested and showed a nodule in the right thyroid gland with radioisotope hyper-uptake suggestive of parathyroid adenoma. The patient subjected to hematological examination (table1).

Table 1: Laboratory data

<table>
<thead>
<tr>
<th>Test</th>
<th>Value</th>
<th>Normal range</th>
<th>Unit</th>
</tr>
</thead>
<tbody>
<tr>
<td>UREA (Bun)</td>
<td>49.8</td>
<td>25-6.4</td>
<td>mmol/L</td>
</tr>
<tr>
<td>Creatinine</td>
<td>408</td>
<td>61.9-114</td>
<td>umol/L</td>
</tr>
<tr>
<td>Uric acid</td>
<td>541</td>
<td>220-547</td>
<td>umol/L</td>
</tr>
<tr>
<td>Calcium</td>
<td>2.23</td>
<td>2.18-2.6</td>
<td>umol/L</td>
</tr>
<tr>
<td>Phosphorus</td>
<td>3.01</td>
<td>0.78-1.65</td>
<td>umol/L</td>
</tr>
<tr>
<td>PTH</td>
<td>352</td>
<td>1.3-9.3</td>
<td>pmol/L</td>
</tr>
<tr>
<td>Alkaline phosphatase</td>
<td>1600</td>
<td>100-450</td>
<td>U/L</td>
</tr>
<tr>
<td>Na</td>
<td>137.4</td>
<td>132 - 146</td>
<td>mmol/L</td>
</tr>
<tr>
<td>K</td>
<td>5.8</td>
<td>3.5 - 5.5</td>
<td>mmol/L</td>
</tr>
</tbody>
</table>

Based on the investigations of this case, Laboratory findings were consistent with severe hyperparathyroidism (markedly elevated PTH level, phosphorus and alkaline phosphatase with nearly normal calcium level). Increased alkaline phosphatase levels were responsible for the diffuse lytic areas in the bone which were observed radiographically. Based on the above clinical, laboratory and radiological findings, final diagnosis of facial Manifestations of ROD was given.

Patient was referred to the Department of Orthopedics for rehabilitation prosthesis and nephrology for regular dialysis procedure.

3. DISCUSSION

Uremic leontiasis ossea refers to the massive thickening of craniofacial bones as a result of CRF [3]. Its clinical presentation includes progressive painless massive enlargement of the jaws, widening of the nares, flattening of the nasal bridge, and increased interdental spacing. In addition to the cosmetic impairment, patients suffer functional impairment, including nerve compromise and potential airway obstruction. These clinical changes can be stabilized or improve mildly after parathyroidectomy [3].

In our case, the pathognomonic facial enlargement is secondary to hyperparathyroidism after long-standing CRF. At the moment of diagnosis, the blood screening showed altered levels of PTH secretion with hyperphosphatemia and normocalcemia. The increase in the secretion of PTH was in response to hypocalcemia and hyperphosphatemia due to CRF.

It's important to note that increased levels of PTH can be detected in blood test analysis prior to clinical features of calcium impaired metabolism, thus it is important to emphasize on patients preventive screening during hemodialysis [4].

Changes in the facial skeleton due to hyperparathyroidism assume 3 known radiographic patterns [5]. Radiographically, the osteolytic lesions have a “salt-and-pepper” appearance, which is the result of mixed osteolytic and sclerotic osseous involvement. The second form resembles fibrous dysplasia, with a classic ground-glass pattern on both conventional films and CT. Unlike true fibrous dysplasia, these findings can be diffuse and generalized, with poor corticomедullary distinction, an imaging not present in fibrous dysplasia. The third pattern is the most uncommon form, and it is present in uremic leontiasis ossea, characterized by significant hypertrophy of the jaws with serpiginous “tunneling” or channeling within the bone and poor visualization of the cortical bone[6].
In our case, the noncontract CT scan showed bony thickening of the palate with low attenuation serpentine “tunneling” extending through the maxilla and mandible. The affected area lacked clearly defined cortical bone with no corticomedullary distinction. Bone biopsy is not particularly helpful in distinguishing uremic leontiasis ossea from fibrous dysplasia and Paget's disease, since both conditions can have very similar histological findings [6]. The differential diagnosis can be assessed based on the combination of clinical, laboratory, and diagnostic imaging findings.

4. CONCLUSION

Uremic leontiasis Ossea diagnosis can be done by the combination of clinical, laboratory, and diagnostic imaging findings. Bone biopsy is not particularly helpful in distinguishing uremic leontiasis ossea from fibrous dysplasia and Paget's disease. Early recognition of incipient uremic leontiasis ossea is essential to prevent progression to severe disfigurement that can result from prolonged untreated secondary hyperparathyroidism.

REFERENCES