Unusually Large Brown tumor of Mandible in a Case of Secondary Hyperparathyroidism Mimicking Cherubism

Abstract

We report here a case of unusually large brown tumor of mandible mimicking cherubism in a patient with secondary hyperparathyroidism (HPT). The patient is a young male with a large head and a protruding jaw with an open mouth appearance. Initial clinical appearance looked like cherubism. However further clinical, biochemical, and radiological evaluation revealed a large brown tumor in a case of prolonged secondary HPT, which was confirmed on histopathology. All of the typical advanced radiological features of HPT were noted, highlighting the severity of progression of the disease. This case emphasizes the need for surveillance of serum calcium levels on routine biochemical investigations as to enable an early diagnosis of HPT. With timely proper management, such cases should be on the decline in the future.

Keywords: *Brown tumor, cherubism, floating teeth, hyperparathyroidism, jaw tumor, parathyroid adenoma*

Alka Ashmita Singhal, Sanjay Saran Baijal, Deepak Sarin, Atul Pathak

Medanta Division of Radiology and Nuclear Medicine, Medanta The Medicity Hospital, Gurugram, Delhi, NCR, India

Introduction

Brown tumor of hyperparathyroidism (HPT), also called osteitis fibroscystica or osteoclastoma[1] arises due to excessive osteoclastic activity as a result of the metabolic bone disorder in HPT.[2] It is seen in about 2% to 3% of cases of HPT,[3-5] incidence reported to be 3% in primary HPT and 1.5%–1.7% in secondary HPT. Earlier diagnosis and effective management of HPT has led to the decrease in its incidence in the developed world. In contrast, the incidence of brown tumors in underdeveloped countries may be as high as 58%–69%.[6]

Case Report

A 41-year-old male from the Middle East, of short stature and short neck [Figure 1], had a grossly enlarged mandible and protruding lower jaw, prognathism, and open mouth appearance. He could not walk on his own and was limited to wheelchair. He was suffering from chronic kidney disease. Bone dexa scan showed severe osteoporosis. On biochemical investigations, serum calcium was increased to 10.2 mg/dl, serum PTH was markedly elevated to 4808.0 pg/mL and 25–OH Vitamin D was 16.3 g/mL (Normal reference values of total serum calcium for adult males 8.9–10.1 mg/dl, serum PTH reference levels for all ages 15–65 pg/ml and for 25–OH Vitamin D levels is 20–50 ng/ml). Clinical diagnosis of secondary HPT was suggested. Plain radiographs [Figure 2a-d] were done. Skull (a) showed typical salt and pepper appearance. Chest (b) showed gross cardiomegaly with mixed lytic and sclerotic areas in both clavicles, scapula, humeri, and multiple ribs. Calcification of the trachea and bronchi was seen. Forearms (c) showed osteopenia with resorption of terminal phalanges of the 1st and 4th fingers. Extensive vascular calcification was noted. Dorso-lumbar spine (d) showed flattened lower thoracic vertebrae, anterior wedging of upper lumbar vertebrae with reduction of intervening disc spaces. Further evaluation by computed tomography for facial bones and paranasal sinuses [Figure 3a-d] showed generalized increased bone density with multiple lytic and sclerotic lesions in all bones of the skull. Gross expansion and marked thinning of the cortex of bilateral maxilla, causing narrowing of the nasopharynx, and nasal cavities were noted. Significant destruction of the alveolar arches in maxilla and mandible was seen giving a "floating appearance

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Address for correspondence:
Dr. Alka Ashmita Singhal, Medanta Division of Radiology and Nuclear Medicine, Medanta The Medicity Hospital, Sector 38, Gurugram, Haryana - 122 001, India. E-mail: dr.alkaasinghal@gmail.com

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to the teeth." Calcific mitral valve leaflet with moderate mitral regurgitation was detected on echocardiography. Tc 99m Sestamibi Parathyroid Scan: [Figure 4a] localized a left inferior parathyroid adenoma. Ultrasound [Figure 4b] in addition to sestamibi revealed all the four enlarged parathyroids as characteristic hypoechoic nodules in their usual location with arc rim vascularity on color Doppler. Right superior parathyroid measuring 24 mm × 9 mm × 18 mm was located posterior to the mid pole of the right thyroid, and right inferior parathyroid measuring 14 mm × 13 mm × 10 mm was located inferior to the lower pole of the right thyroid gland. Left superior parathyroid measuring 11 mm × 6 mm × 5 mm was located posterior to left mid thyroid and left inferior parathyroid measuring 16 × 13 mm × 14 mm was located inferior to the lower pole of the left thyroid. Evaluation with radionuclide bone scan (20 mCiof Tc-99m MDP) [Figure 5] showed focal abnormal uptake in the grossly enlarged mandible and maxilla. Increased uptake was also noted in calvarium.

Histopathology

Bone biopsy was performed from the right maxillary bone via the oral route and a single gray white tissue piece measuring 17 mm × 8 mm × 5 mm was received, trisected, and all processed. Microscopic examination [Figure 6a and b] showed fibro-collagenous tissue admixed with irregular spicules of woven trabecular bone and multiple multinucleated osteoclastic type giant cells. Foci of calcification noted. No evidence of dysplasia or malignancy. Histopathological findings[7] were consistent with brown tumor syndrome.

Management

The patient was planned for parathyroidectomy, but unfortunately, the patient expired before surgery.

Figure 1: Patient photo showing enlarged head and face with prognathism, open mouth and short neck

Discussion

Brown tumors[8-10] are nonneoplastic osteolytic lesions in the bones due to excessive osteoclastic activity as a part of the metabolic abnormality in HPT. Any bone may be involved, though common locations are mandible, maxilla, palate, nasal cavity, paranasal sinuses, orbit, and temporal bones. Ribs, clavicle, spine, extremities, and pelvic girdle bones are also involved. There is demineralization, altered bone trabecular pattern and replacement by loose connective tissue or woven bone. With craniofacial brown tumors, this reduced density (radiolucency) of the jaw bones in contrast to the dense teeth gives rise to "floating teeth" appearance. Large mandibular and maxillary brown tumors[11] can cause facial disfigurement and compromise on the normal routine functions of chewing, talking, and breathing. The usual management is surgical resection[12-14] of the parathyroid adenoma and the bone lesions regress thereafter. Surgery for bone lesions is rarely needed.

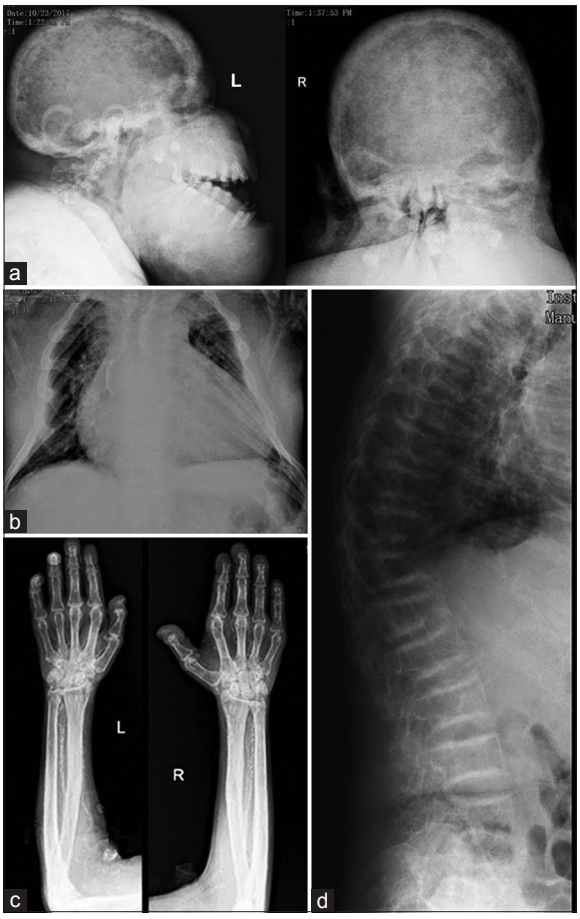

Figure 2: Skull radiograph (a) showing typical salt and pepper skull, enlarged maxilla and mandible with prognathism. Chest radiograph (b) with cardiomegaly, mixed lytic and sclerotic areas in clavicles, scapula, humerus and ribs. Calcification of trachea and bronchi. Forearms radiograph (c) showing resorption of terminal phalanges of 1st and 4th fingers and extensive vascular calcification. Lateral spine radiograph (d) showing generalized osteopenia, flattened lower thoracic vertebrae, anterior wedging of upper lumbar vertebrae with reduction of intervening disc spaces

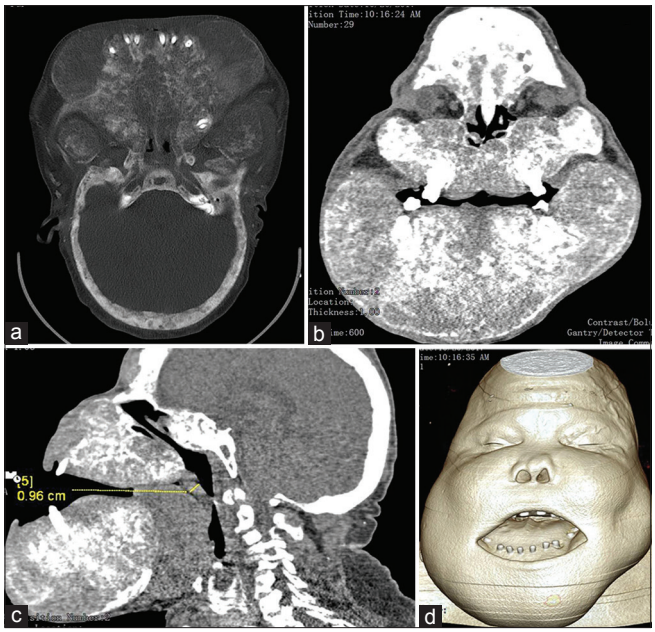

Figure 3: (a-d) Computed tomography Facial Bones and PNS: showing thickened skull vault with loss of definition of inner and outer tables, increased bone density with multiple lytic and sclerotic lesions of skull vault, clivus, facial bones and alveolar arch. Gross expansion and thinning of the cortex of bilateral maxilla, greater and lesser wings of sphenoid, pterygoid plates and mandible. Resorption of paranasal sinuses and mastoids. Expanded mandible displacing the tongue posteriorly causing narrowing of the oropharynx and hypopharynx. Destruction of the alveolar arches giving "floating teeth appearance"

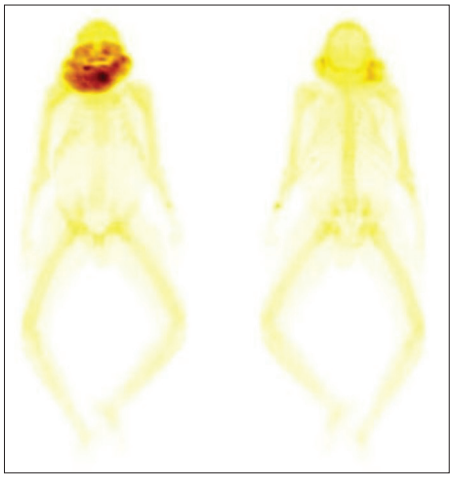

Figure 5: Radionuclide Bone Scan (20mCiof Tc-99m MDP) showing focal abnormal uptake in the grossly enlarged mandible and maxilla. Increased uptake also noted in calvarium

Cherubism

With the enlarged mandible and floating teeth appearance and short stature, cherubism was initially considered as a differential diagnosis. Cherubism[15] is a childhood disorder characterized by abnormal bone tissue in the lower part of the face. Both upper and lower jaws are enlarged due to the replacement of the bone by cysts. It is painless and causes chubby cheek appearance and may cause dentition, swallowing, speech, and breathing problems. Enlargement of jaw stabilizes by puberty. It may be a part of genetic syndromes as Noonan syndrome, or fragile X syndrome.

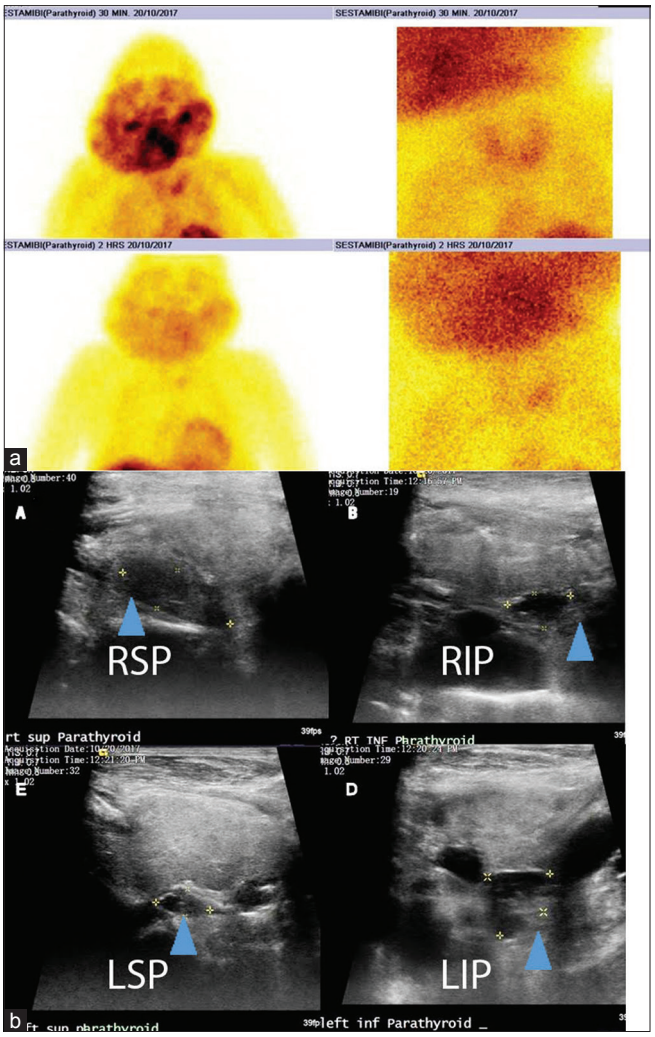

Figure 4: Tc 99m sestamibi parathyroid Scan (a): Increased uptake at the lower aspect of left lobe of thyroid which persists on the delayed image at 2 hours while the thyroid gland shows normal washout, suggestive of left inferior parathyroid. Increased uptake is noted in the maxilla and the mandible. Ultrasound (b): All four enlarged parathyroid nodules seen in usual locations. Right superior parathyroid (RSP) 24 mm × 9 mm × 18 mm behind right mid thyroid. Right inferior-14 mm × 13 mm × 10 mm at right thyroid lower pole. Left superior-11 mm × 6 mm × 5 mm–behind left mid thyroid. Left inferior-16 mm × 13 mm × 14 mm at left thyroid lower pole

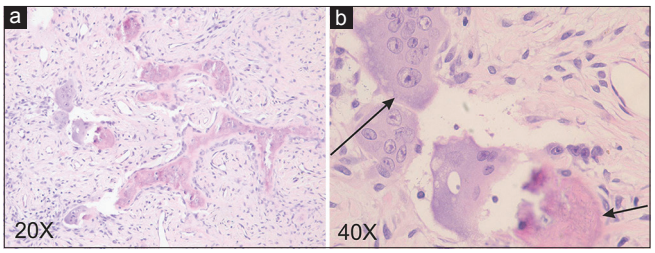

Figure 6: Histopathological photomicrograph (a) - ×20 and (b) - ×40 showing fibrocollagenous tissue containing irregular spicules of woven trabecular bone and numerous osteoclasts in the areas of bone resorption. Long Arrow indicates multinucleate giant cell. Short arrow indicates bone

Conclusion

Prolonged HPT with large brown tumors and the characteristic severe advanced radiological features of HPT are still being seen in developing countries where there is lack of medical care and the entity is often undiagnosed and left untreated. Therefore, it is imperative to develop a protocol for its early detection. To increase the surveillance for hypercalcemia serum calcium levels may be included in basic routine biochemical investigations. Further, followed by serum parathyroid hormone levels in cases of hypercalcemia, it will help in making an early diagnosis of asymptomatic HPT. This needs to be emphasized and coordinated by all the involved health-care professionals, clinicians, dentists, gastroenterologists, nephrologists, orthopedicians, endocrinologists, oncologists, surgeons, and anesthetists to curtail such advanced cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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