CASE REPORT

Tertiary Hyperparathyroidism Inducing Brown Tumour: A Rare Case

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ABSTRACT

Brown tumours (BTs), also known as osteitis fibrosa cystica, are non-neoplastic giant cell lesions which appear in the advanced stage of hyperparathyroidism. Patients with giant cell lesions should be screened for serum calcium, parathyroid hormone (PTH) and alkaline phosphatase. The BT may affect any part of the skeleton but are frequently found in long bones, pelvic girdle, clavicle, ribs and the mandible. The authors present a rare case of BT in tertiary hyperparathyroidism (THP) involving bilateral maxilla, left maxillary sinus, lateral wall of the nose, floor of the orbit and bilateral mandible in a 28-year-old male patient.

INTRODUCTION

Brown tumours (BTs) also known as osteitis fibrosa cystica are non-neoplastic giant cell lesions that appears in advanced stage of hyperparathyroidism.1-4 BT was reported in 1891.5-8 The reported prevalence of BT is 0.1% and its incidence is 4.5% in primary hyperparathyroidism6 and 1.5-1.7% in secondary hyperparathyroidism.2,6,7 The reported incidence of tertiary hyperparathyroidism (THP) is <30% at 2 years post transplantation and about 17% at 4 years post transplantation, but the true incidence might be lower (5.6% or less).9

BT represent focal bone lytic lesions often developing at multiple sites.3 BT may affect any part of skeleton but are frequently found in long bones, pelvic girdle, clavicle, ribs and the mandible. Tumours involving the maxilla and orbit are quite rare.2-4 This disease can manifest itself at any age, but is more common over 50 years of age, and is three times more common in women.2,3,6,10 Solitary tumour may resemble a central giant cell granuloma or an aneurysmal bone cyst.3,7 The radiographic findings can mimic bone malignancy, while the synchronous involvement of multiple skeletal segments can be interpreted as diffuse metastatic disease.1 It is interesting that the histological appearance of the BT is identical to that of the central giant cell granuloma. Therefore, patient with giant cell lesions should be screened for serum calcium, parathyroid hormone (PTH) and alkaline phosphatase.3

In this article we present a rare case of BT in THP involving bilateral maxilla, left maxillary sinus, left lateral wall of the nose, left floor of the orbit and bilateral mandible in a young male patient.

CASE REPORT

A 28-year-old male patient reported with a complaint of bilateral painful swelling in maxilla and mandible. The swelling was initially small and asymptomatic which gradually increased in size over a period of 6 months in maxilla when he first noticed it on the palate and after 4 months he noticed a similar type of swelling in mandibular right premolar region with mobility of teeth in the same region and maxillary anterior region. The swelling was painful since 20 days. There was history of intense fatigue, weight loss (of 4 kg) decreased appetite and bone pain, most prominent in the knees since 2 months. He also complained of difficulty in breathing from the left nostril. His gait was abnormal. He was poorly built and had pallor. There was no family history of endocrinopathies or bone disease. On examination there was mild proptosis of the left eye and bilateral swelling noticed in maxilla and mandible leading to facial asymmetry (Fig. 1a). The swelling was tender and bony-hard on palpation situated in the labial and palatal aspect in the maxillary anterior region bilaterally from 14 to 22 and right mandibular premolar region labially extending from 41 to 45 obliterating the labial vestibule (Fig. 1b), which exhibited crepitus in certain regions. The adjacent teeth grade I mobile and tender on percussion. The left nasal passage was completely obstructed.

PNS X ray view revealed radiopacity in left maxillary sinus (Fig. 2). CBCT scan showed expansile, multilocular, well defined, corticated radiolucency extending mesiodistally from 17 to 27. It involved the maxillary sinus especially on the left side, and the alveolar ridge in the apical one-third area of the roots. There was destruction of the medial, lateral, anterior and floor of the maxillary sinus and the left lateral wall of nose and lateral part of floor of left bony orbit.

KEYWORDS hyperparathyroidism, osteitis fibrosa cystica, facial bones

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Complete opacification of the maxillary sinus was seen, with discontinuity in the floor in 16, 17 region. There was an expansion of the buccal and palatal cortical plates in maxillary anterior 14, 15 and 26, 27 regions. Also there was an external root resorption in relation to 11, 12 and 21 (Fig. 3a). In the mandibular section, a fairly defined, mixed, non corticated, radiolucent and sclerotic areas involving the entire body of mandible on both sides were seen. There was thinning of the inferior border of mandible and loss of lingual cortical plate in 36–37 regions with external root resorption of roots of both the teeth (Fig. 3b).

The differential diagnosis considered at the time included central giant cell granuloma, aneurysmal bone cyst and “brown tumour” of hyperparathyroidism.

Routine blood investigations showed anaemia with Hb of 9.7 gm/dL, hypercalcemia with serum calcium level >15 mg/dl, vitamin D deficiency with 5 ng/ml. Bence Jones protein test was found to be positive.

Incisional biopsy from the mass in the left maxillary sinus and right mandibular buccal aspect was done under local anaesthesia. The histopathology report showed giant cell granuloma. This report, along with the blood chemistry report indicating hypercalcemia, prompted referring the patient for evaluation of THP. Further laboratory investigations conducted by the endocrinologist were as follows: serum albumin 4.1 gm%, alkaline phosphatase 993 IU/L (33-96), and very high PTH levels, 1198 pg/ml (normal value 15–65 pg/ml), confirming THP. The renal function tests showed blood urea 45mg% (15–45 mg%), serum creatinine 1.3 mg% (0.6–1.4 mg%), serum protein 7.4g% (6–8 g%), serum globulin 3.3g% (2–3.5 g%) which were within normal limits.

The patient was advised MRI and a parathyroid scan to rule out for adenoma of parathyroid gland by the physician. The gland was normal and he was advised oral medications for the raised parathyroid levels.

Later, the patient had a fall on the left knee, following a mere push from an unknown person, on the left arm and immediate left leg and left hand X-rays revealed a complete oblique minimally displaced fracture of proximal shaft of femur. Also there was a displaced complete comminuted fracture of left humerus and was then surgically operated for the fractured humerus and femur (Fig. 4a, b).

The patient was advised to take vitamin D supplements and have a regular follow up.

DISCUSSION

Von Recklinghausen was the first to describe hyperparathyroidism (HPT) as a bone disease in 1891. Askanazy, in 1904, was the first to describe a patient with a parathyroid tumour and osteitis fibrosa cystica. However, he did not correlate these two entities. In 1962, Mandle operated on a parathyroid tumor in a patient with hypercalcemia and radiological changes of osteitis fibrosa cystica, and demonstrated post-operative regression of

![Image](image_url)
Tertiary hyperparathyroidism inducing BT

Fig. 3  a: CBCT scan of maxilla, showing expansile, multilocular, well defined, corticated radiolucency extending mesiodistally from 17 to 27. b: CBCT scan of mandible, showing defined, mixed, non corticated radiolucent and sclerotic areas involving the entire body of the mandible on both the sides with thinning of the inferior border of the mandible

Fig. 4  a: Comminuted fracture of the left humerus. b: Complete, oblique, minimally displaced fracture of proximal shaft of the left femur.

the bone disease and biochemical abnormalities.\(^5,8\) HPT may occur in one of three clinical forms: primary, secondary or tertiary. In all instances the disease is characterised by increase in PTH level and mobilisation of calcium from bone. Primary parathyroid hyperplasia, parathyroid adenoma or parathyroid carcinoma can result in primary HPT. Secondary HPT results from a compensatory mechanism due to the primary condition producing hypocalcemia such as rickets, osteomalacia, pregnancy or chronic renal insufficiency. THP may occur after long-standing secondary HPT. Increased level of PTH causes bone resorption that result in hypercalcaemia.\(^2,5,7,8,11\)
A fourth type of HPT, (ectopic HPT) as described by Guimarães et al is thought to arise from increased PTH levels synthesised in patients with malignant diseases. In our case, it was noticed that the increased levels of parathyroid, serum calcium and serum alkaline phosphatase levels was due to long standing secondary hyperparathyroidism which was induced by vitamin D deficiency.

The name ‘brown tumour’ for bony lesions seen in HPT was first coined by Jaffe. The term “tumour” in BT is a misnomer. It’s also known as Osteoclastoma. The term “brown tumor” is derived from the characteristic appearance of brownish material within the cystic lesion, which is caused by the vascularity, haemorrhage, and deposits of haemosiderin. BTs are non-neoplastic lesions representing a reparative cellular phenomenon that involves areas with intense bone re-absorption due to the effect of high circulating PTH levels. It is considered as a kind of giant cell lesion that presents as an osteolytic lesion of the bone. Giant cells refill the osteolytic defect as fibroblasts and can deform the bone profile, mimic a neoplasm or cause pathological fractures. In present case patient had a complete oblique minimally displaced fracture of proximal shaft of femur and a displaced complete comminuted fracture of left humerus on a mere push suggestive of fracture.

BT may affect any skeleton but are frequently found in long bones, pelvic girdle, clavicle, ribs and the mandible. The tumours involving the maxilla and orbit are quite rare. In our case, it was involving bilateral maxilla, left maxillary sinus, left lateral wall of nose, floor of the orbital and bilateral mandible. The disease can manifest itself at any age, but is more common over 50 years of age, and is three times more common in women unlike present case who was a young male of 28 years.

BT appears as a well-defined unilocular or multilocular radiolucency. Ashrafi described their radiographic appearance as well-defined lytic lesions with little reactive bone formation, cortical thinning or expansion. Langland et al. described them to be monocystic or fibrosa cystic. In the present report, CT revealed expansile, multilocular, well defined, corticated radiolucency in maxilla with opacification of the maxillary sinus and fairly defined, mixed, non corticated, radiolucent and sclerotic areas in the mandible.

Treatment of BT should be aimed at treating the HPT state. Many lesions spontaneously regress once the etiology is removed. Regression of BT can also be achieved by medical treatment using intravenous calcidiol, calcitriol and vitamin D supplements. Treatment with conservative surgical debridement and replacement therapy has been reported to yield satisfactory results in 6 months.

To the best of our knowledge, a case of tertiary HPT due to vitamin D deficiency is very rare. This case illustrates that BT of tertiary HPT can develop in vitamin D deficiency and there is a response to vitamin D supplements. The patient's recovery after supplements is progressive.

CONCLUSION

BTs rarely involve the maxilla, lateral wall of nose, floor of the orbit along with mandible in association with tertiary HPT in young male patient. It is therefore necessary to remind practitioners for thorough diagnostic workup for all giant cell lesions in maxillofacial region. A parathyroid estimation, calcium, phosphorous and alkaline phosphatase levels should be made a mandatory investigation in all cases of giant cell lesions.

REFERENCES


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