DOI: 10.7860/JCDR/2014/9647.4727

Case Report

Dentistry Section

Brown Tumour of Posterior Maxilla Associated with Polydactyly, Syndactyly and Cardiac Anomalies: A Unique Case Report

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ABSTRACT

Brown tumour [BT] is an uncommon, non neoplastic complication of Hyperparathyroidism [HPT]. Skeletal changes are the main consequences of this endocrine condition. HPT manifest in three categories. Primary HPT is due to hyperfunction of one or more parathyroid gland, causing an increase of parathyroid hormone secretion resulting in hypercalcemia. Secondary HPT occurs due to chronic renal failure, decreased vitamin D production or with hypocalcemia. Tertiary HPT occurs when the parathyroid activity turns autonomous and excessive, leading to hypercalcemia. BT occurs frequently in the mandible than the maxilla. They are more common in women aged over 50y and majority of BT are asymptomatic. Radiographically, it appears as well defined radioluceny. The gross specimen usually shows a brown or reddish-brown colour. We report a relatively rare combination of maxillary posterior BT as a clinical manifestation of secondary HPT due to vitamin D deficiency with polydactyly, syndactyly and cardiac anomalies.

Keywords: Brown tumours, Parathyroid hormone, Secondary hyperparathyroidism, Vitamin D deficiency

CASE REPORT

A 17-year-old male patient reported with a complaint of an asymptomatic unilateral swelling in the left maxillary posterior region of six months duration. It was initially small and gradually increased to its presenting size of approximately 5 x 4 cm. Intra oral examination revealed a well defined, bright pink coloured swelling with buccal and palatal cortices expanded [Table/Fig-1]. Oral hygiene status was poor with multiple decayed teeth. The patient also complained of general weakness and difficulty in doing routine work. On general examination he was moderately built and nourished with all vital signs within normal limits. Syndactyly in both right and left great toe and polydactyly in left hand was noticed.

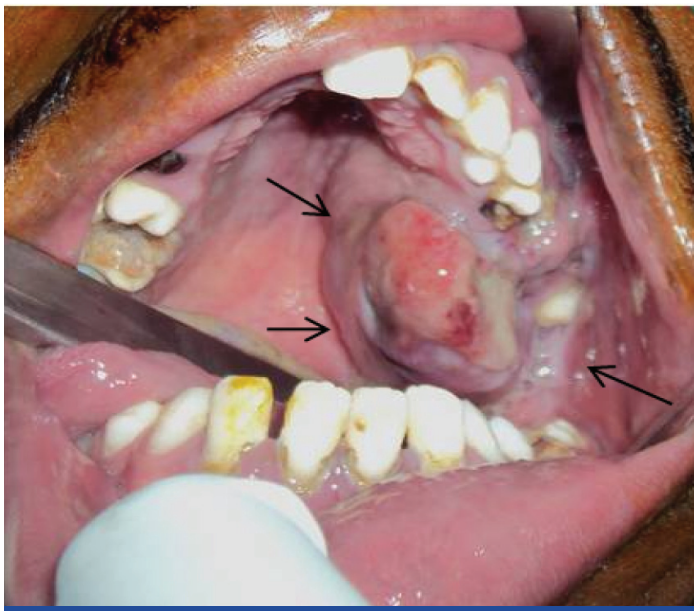
The Intraoral and Panoramic radiograph showed a well-defined radiolucency of 3x2 cm involving posterior left maxillary region. Teeth in the affected region showed displacement. Floor of left maxillary sinus was displaced in upward direction. First molar (26) was found missing. Tapered roots in 25, 37, 31,32,41,42, 43 and 44 were noted. Generalized alteration of trabecular bone in maxillary and mandibular jaws with loss of lamina dura was noticed. Cortical borders in mandible and maxillary sinuses appeared normal [Table/Fig-2]. Hand-wrist and Foot radiographs revealed polydactyly on left hand and Syndactyly on right and left great toe [Table/Fig-3]. Echocardiography showed evidence of single atrium. Full skeletal investigation ruled out the presence of any bony lesions in other bones. An incisional biopsy was taken and histopathological examination of specimen revealed a cellular connective tissue comprising spindle cells and osteoclast-like giant cells with osteoblastic rimming and new bone formation. Numerous foci of extravasated RBC's with hemosiderin pigments were seen [Table/Fig-4] Histological diagnosis of a giant cell lesion was considered and patient was subjected for PTH estimation, renal function tests, and complete blood investigation.

Haemtological analysis demonstrated an elevated Paratharome level of 165.7 pg/mL (Normal =12 to 72 pg/mL), Serum calcium level of 9.3mg/dL (Normal = 8.4 to 10.4 mg/dL), Ionized calcium level of 1.23mmol/L (Normal =1.10 to 1.40 mmol/L), serum phosphate level of 2.2 mg/dL (Normal = 2.5 to 4.8 mg/dL), reduced serum 25 OH vitamin D level at < 6 ng/mL (Normal 10 to 60 ng/mL).

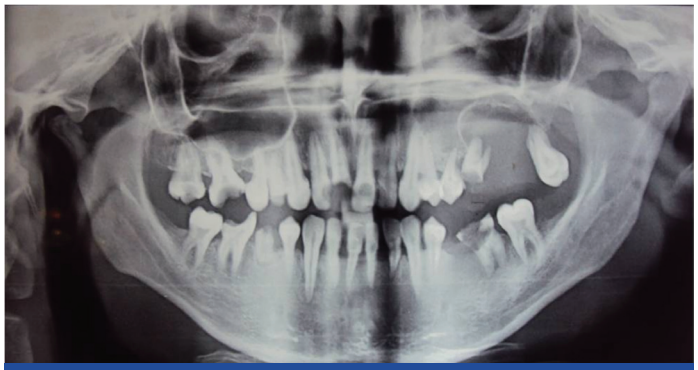
Heamogram, coagulation test, glycosylated hemoglobin and renal function tests were within the normal limits. A final diagnosis of brown tumour secondary to a deficiency in vitamin D was arrived based on the hematological data and characteristic histopathologic findings. Complete resection of the tumour was performed under general anesthesia [Table/Fig-5] Grossly, the specimen was dark reddish brown in colour. The microscopic findings were similar to the incisional biopsy specimen

The patient was prescribed regular calcium supplements and 30000 units / ml of vitamin D25 injections by endocrinologist, which slowly increased the vitamin D level to 24 ng/mL. He was also advised to regularly consume dairy products and get exposure to sunlight.

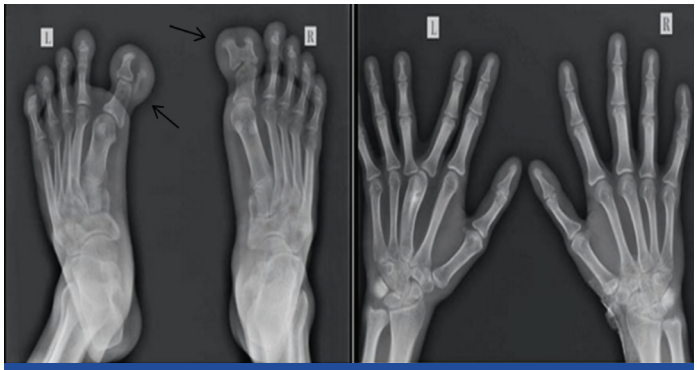
Patient was followed-up after 10 months with all investigations repeated. The lesion was completely healed and all test values were within the normal range. The patient is continuous surveillance for the past three years and has not exhibited any signs or symptoms of recurrence.



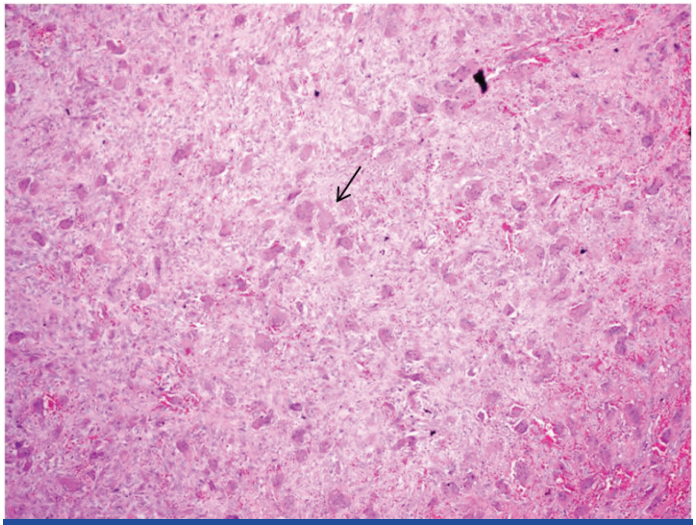
[Table/Fig-1]: Well defined swelling in left maxillary posterior region



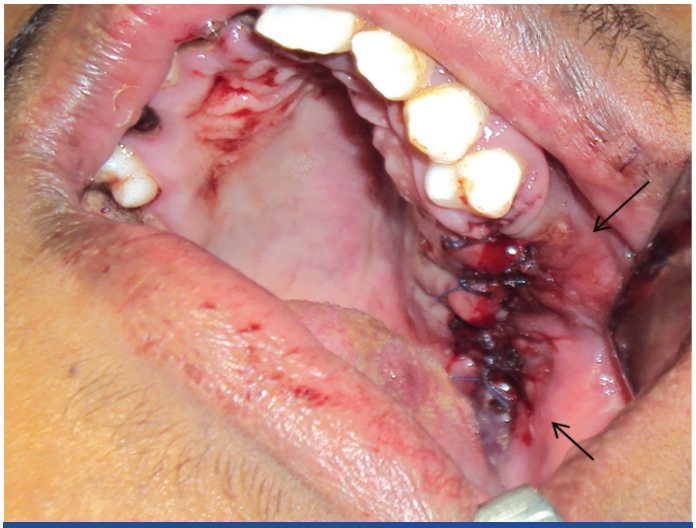
[Table/Fig-2]: Panoramic radiograph showing well-defined radiolucency of 3x2 cm involving posterior left maxillary region



[Table/Fig-3]: Hand - wrist and foot radiograph reveals polydactyly on left hand and syndactyly on right and left great toe



[Table/Fig-4]: H and E (4x) stained photomicrograph showing multinucleated giant cells in cellular stroma and hemorrhagic areas



[Table/Fig-5]: Postoperative photographs showing complete excision of the swelling

DISCUSSION

BT are non-neoplastic giant cell lesions associated with primary or secondary HPT. In the past, bone lesions were reported to occur in 80% to 90% of patients with HPT. But in the last few years these rates have been declined to 10% to 15%. This has been attributed to the early diagnosis and successful treatment of this disease. These lesions may be single or multiple within bone and very rarely affects the soft tissues [1]. BT may affect any skeleton but are frequently found in long bones, pelvic girdle, clavicle, ribs and the mandible. Tumours involving the maxilla are quite rare [2,3]. The brown colour of the lesion is due to increased vascularity, hemorrhage and deposits of hemosiderin. They have variably defined margins and may produce cortical expansion. Solitary tumour may resemble a central giant cell granuloma or an aneurysmal bone cyst. 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, PTH, and alkaline phosphatase [4]. This papers aims to highlight a rare case of BT of posterior maxilla in a 17-year-old male patient as a consequence of vitamin D deficiency.

The term “tumour” in BT is a misnomer. It’s also known as Osteoclastoma which represents cellular reparative processes with increased granulation tissue and rich vascularity. BT consists of multiple giant cells mixed with stromal cells and matrix. The cystic spaces are filled by hemorrhage, hemosiderin and hypervascularity lead to the brownish colour, which is responsible for the name [5].

Prevalence of BT is about 0.1%, more common in persons older than 50 y and more frequent in mandible. In contrast, our patient’s lesion was seen in maxillary posterior region which is very rare and moreover age of occurrence was in second decade. The true prevalence of BT is actually higher than reported in the literature. The likely explanation is that in most patients, the jaw lesions are never diagnosed and will spontaneously disappear when the PTH and calcium levels are corrected [6,7].

Secondary HPT is due to chronic kidney diseases and vitamin D deficiency. Vitamin D deficiency is due to a dietary deficiency or lack of sunlight exposure. It is caused by conditions that result in little exposure to sunlight, such as living in northern latitudes, dark skin, infants or elders having less chance to go outside, and covering one’s face and body mainly due to religious reasons. Particularly, women may acquire vitamin D deficiency, even though they live in a sunny climate [8,9].

The majority patients reported with maxillary lesions present as hard, clearly visible and palpable swellings, with occasional pain [10,11]. Our case presented with an exuberant swelling in left maxillary alveolar ridge. BT presenting as proliferative mass is rare except only two cases reported as exophytic masses in mandible. As per our knowledge ours is the first proliferative case reported in maxillary region.

Radiologically, BT present as well-circumscribed unilocular or multilocular osteolytic lesions. In the mandible, cortical bone is usually expanded and thinned. BT of jaws results in root resorption and loss of lamina dura (40% of cases) [1]. The density of jaws is decreased due to generalized demineralization of the medullary bones along with change in trabecule pattern giving a mixed radiopaque-radiolucent appearance. Osteitis fibrosa cystica is a late manifestation of severe HPT.

Our case had a unilocular radiolucency, extending from maxillary left first molar to maxillary left third molar and displacement of roots

of teeth and loss of lamina dura in all teeth. Histological features alone cannot establish a diagnosis of a BT. However, a clinical history of common skeletal involvement, elevated serum calcium and parathyroid hormone levels helps in diagnosis [11].

Treatment of BT should be aimed at treating the HPT state. Regression or resolution of the tumour occur following vitamin D supplements. Many lesions spontaneously regress once the aetiology 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 [9].

To the best of our knowledge, only one pediatric maxillofacial case and one case in the metacarpal bone with brown tumours secondary to dietary vitamin D deficiency and inadequate sunlight exposure have been reported [12]. This case illustrates that BT of secondary HPT can develop in vitamin D deficiency and there is a response to vitamin D supplements.

CONCLUSION

Brown tumours of the jaw rarely involve the maxilla in association with secondary HPT. It is therefore necessary to remind practitioners for thorough diagnostic workup for all giant cell lesions in maxillofacial region. A parathyroid estimation, calcium and phosphorous, alkaline phosphatase levels should be made a mandatory investigation in all cases of giant cell lesions.

ACKNOWLEDGEMENT

My heartful gratitude to Dr. Modak Abdulfajar, MBBS, D Ortho [Modak Orthopedic hospital and Fracture center, Kolhapur] for his support and cooperation. We also wish to acknowledge Dr Ajay G Nayak, Associate Professor, Department of Oral Medicine and Radiology, School of Dental Sciences, who inspired to write this article.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Apr 15, 2014**
Date of Peer Review: **Jun 01, 2014**
Date of Acceptance: **Jun 19, 2014**
Date of Publishing: **Aug 20, 2014**