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A RARE CASE OF MAXILLARY BROWN TUMOUR AS PRIMARY PRESENTATION OF THE PARATHYROID CARCINOMA.

Oncology

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ABSTRACT

Background: Brown tumours are expansile osteolytic lesions of bone, occurring in Hyperparathyroidism. Brown tumours occur most commonly in ribs, clavicle, long bones and pelvis and are uncommon in other facial bones except mandible. Other facial bones are rarely affected. Brown tumors are due to the direct effect of the parathyroid hormone. Brown tumors occur more with primary hyperparathyroidism than secondary. However, they are reported more in secondary hyperparathyroidism. In primary hyperparathyroidism, a parathyroid adenoma is a cause in 81% while other causes include hyperplasia in 15% and parathyroid carcinoma only in 4%. We present a case report of maxillary Brown tumor due to parathyroid carcinoma in an elderly male patient.

Case Report: A 67-year-old male presented with right maxillary swelling increasing in size for the last few months associated with ipsilateral nasal block and right eye epiphora. The contrast CT scan of paranasal sinuses and neck revealed a large expansile right maxillary tumor aggressively eroding maxillary wall with extension into the orbital floor, ethmoid, sphenoid sinuses, nasal cavity, and oral cavity with the erosion of hard palate and soft tissue extension to subcutaneous Plane. A three cm sized soft tissue density lesion was also noted posterior to the right thyroid lobe in CT sections of the neck.

Blood profile was normal except extremely high serum parathormone and calcium as well as mildly elevated serum creatinine (S. PTH 3437 pg./ml. S. Ca. 19 mg%. S. Creatinine 1.77mg%.) Ultrasonography of the abdomen also revealed calcification in the renal medulla.

Right lower parathyroidectomy was done with the frozen section as well as the Intraoperative Rapid PTH assay. The PTH level was reduced by 90 percent of the original value. The final histopathology was suggestive of parathyroid carcinoma.

Summary: The patient was under regular surveillance, as the maxillary tumor was under remittance after the resection of parathyroid carcinoma. Parathyroid carcinoma is a very rare tumor and involvement of maxillary bone due to primary hyperparathyroidism due to parathyroid carcinoma is also uncommon.

KEYWORDS

Maxillary, Brown tumor, Parathyroid Carcinoma, Hyperparathyroidism, Hypercalcemia

1.INTRODUCTION

Brown tumours are expansile osteolytic lesions of bone, occurring in Hyperparathyroidism. Brown tumours occur most commonly in ribs, clavicle, long bones and pelvis and are uncommon in other facial bones except mandible.

Brown tumors are due to the direct effect of the parathyroid hormone. Brown tumors occur more with primary hyperparathyroidism than secondary. However, they are reported more in secondary hyperparathyroidism. In primary hyperparathyroidism, a parathyroid adenoma is a cause in 81% while other causes include hyperplasia in 15% and parathyroid carcinoma only in 4%. (Sandelin et al.)[1]

The female: male ratio is 1:1 in Parathyroid carcinoma (PC) in contrast to adenoma in which the ratio is 3-4: 1. Brown tumors have been reported to occur in approximately 4.5% of patients with primary hyperparathyroidism (HPT), but they very rarely are the presenting feature[2](Levin KE et al.). The presentation of parathyroid carcinoma with a brown tumor of the mandible or maxilla has been scarcely described in previous case reports

We present a case report of maxillary Brown tumor due to parathyroid carcinoma in an elderly male patient.

2.A Case Report

A 67-year-old male patient presented with right-sided face swelling, which was gradually progressing in size for the last 3 months associated without complaints of pain. He had also suffered from the stuffiness of the right nose and the watering of eyes on the right side. There was no significant family history, past history, or comorbidities. Physical examination revealed a hard and fixed skeletal structure mass in the right maxilla (Figure 1). Examination of the oral cavity, nasal cavity, and neck was unremarkable.

Figure-1 Before treatment pictures of the patient showing right-sided facial swelling

CT scan of Head & Neck showed a large expansile right maxillary tumor aggressively eroding maxillary wall with extension into the orbital floor, right ethmoid sphenoid sinuses, nasal cavity, and oral cavity. Erosion of the hard palate was also noted with soft tissue extension to the subcutaneous plane. (Figure 2.a, b) There was a large more than 3 cm mass posterior to right thyroid lobe on CT Sections of neck (Figure2.c, d) Punch biopsy of the right maxillary lesion was done via trans nasal route and histopathology of the same was suggestive of giant cell tumor.

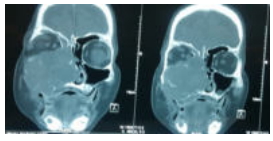

Figure 2 A- Coronal CT images showing Right-sided maxillary brown tumor

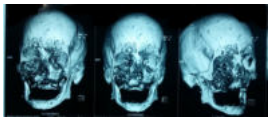

Figure 2 B- 3D Reconstruction of CT images showing erosion of hard palate and right maxilla

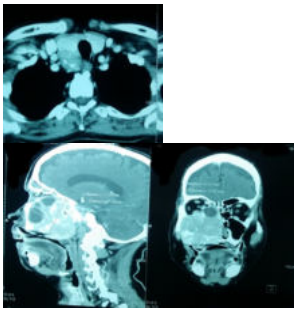

Figure 2-C, 2-D Axial images of CT Neck showing the parathyroid tumor

He had no symptoms suggestive of hoarseness of voice, dysphagia or dyspnoea, recent polydipsia, polyuria, weakness, fatigue, weight loss, palpitations, and recurrent constipation/diarrhoea. Renal sonography revealed increased echogenicity of both cortices and bilateral medullar calcinosis.

S. Parathyroid hormone (S.PTH) was 3437 pg./ml, Calcium was 19 mg% and S. Creatinine was 1.77 mg%. Above mentioned investigations were strongly raised suspicion for parathyroid carcinoma. The patient underwent Right Lower parathyroidectomy with a frozen section biopsy of the gland. During the operation, a capsular mass of 2.5 cm x 2.5 cm x 3 cm attached to the surrounding tissue in the posterior part of the right thyroid lobe was detected (Figure 3) and sent for an intra-operative frozen section. Intra-operative Rapid PTH assay was done after 20 minutes of excision, which was 256 pg/ml. The intraoperative frozen section of the gland revealed parathyroid carcinoma.

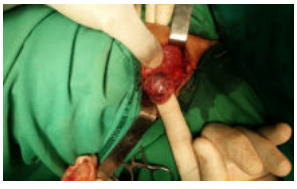

Figure 3- Intraoperative picture showing parathyroid tumor

The final paraffin histopathological sections show polymorphic hyperchromatic cells with capsular and vascular invasion, as well as fibrotic tissue, characteristic of Parathyroid carcinoma (Figure 4 a, b).

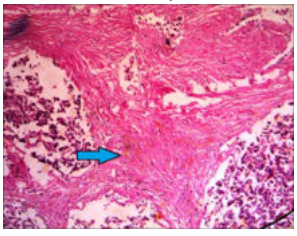

Figure 4 A- Histopathological image showing broad fibrous septa of parathyroid carcinoma.

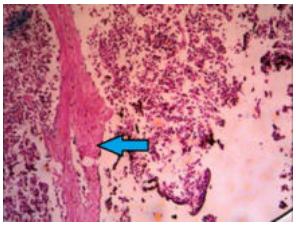

Figure 4 B- Histopathological image showing capsular invasion characteristic of parathyroid carcinoma.

Symptomatic hypocalcaemia following the operation was treated with calcium gluconate.

Pt. was under surveillance as the maxillary Brown tumor was undergoing remittance after parathyroid tumor removal.

Examination at one year post operation showed resolution of the all-maxillary symptoms, a reduction in the size of the brown tumor, and no evidence of recurrence on neck ultrasound. Six years postoperatively he remained asymptomatic, with regression of the mass, as expected. There was no evidence of recurrence of the parathyroid carcinoma, and the PTH level was 41.44 ng/l (normal range 15 to 65 ng/l). Subsequent renal ultrasonography revealed two benign cysts in the right kidney

3.DISCUSSION

Brown tumor is more common in appendicular rather than an axial skeleton. As reported in our case report brown tumor is present in the maxilla, which is less common than the mandible. As reported before, it is unusual for a patient with primary hyperparathyroidism (HPT) to present primarily with a brown tumor, as well as an underlying cause of the primary HPT is parathyroid carcinoma rather than parathyroid adenoma, which is very rare. The reported prevalence of brown tumors has decreased to <0.1%[3](Miyakoshi et al.-2007). Owing to the standard of medical care and screening in developed countries, it is increasingly rare to find accompanying bone disease in primary hyperparathyroidism. 5 Reported cases of extensive multiple brown tumors, some of which are mimicking invasive malignancy, are due to hyperparathyroidism from parathyroid adenoma[4](Hoshi M et al.). There are only a few cases reported where brown tumors are caused by parathyroid carcinoma—two cases of mandibular brown tumor and two cases of multiple brown tumors in the lower extremities[5-8](Pahlavan PS et al-2006, Masson EA-1993 Pai M-1997 Radulescu D-2014).

This exceptional case of Parathyroid carcinoma in combination with a brown tumor of the maxilla is the fifth known case worldwide. Brown tumors, a giant cell granuloma taking place in osteitis fibrosis cystica, can result from both primary and secondary hyperparathyroidism. These tumors are well-defined vascular lesions and might comprise necrotic centres and hemosiderin deposition[9](Holmes EC et al.). These lesions are due to a disparity of osteoblastic and osteoclastic activity and altering bone metabolism, causing bone resorption and fibrous replacement[10](Wiseman SM et al).

Brown tumors are both histologically and radiologically similar to other giant cell lesions including giant cell tumor, giant cell reparative granuloma, aneurismal bone cyst, and cherubim 10(Wiseman SM et al). The tumors can be recognized by checking the PTH and serum calcium levels in combination with clinical conclusions.

The level of hypercalcemia in PC is significantly higher than in adenoma, such as in this patient whose calcium is more than 19 mg/dl. Based on the Schantz and Castleman diagnostic criteria, fibrotic capsular and vascular invasion are key features of PC.

Standard imaging techniques, namely neck ultrasound and MIBI (99m Tc-sestamibi scintigraphy), and, in selected cases, computed tomography and magnetic resonance imaging, although sensitive, have a limited specificity because they cannot accurately distinguish preoperatively a PC from a benign parathyroid tumor unless signs of local invasion are evident. The accuracy of ultrasound, computed tomography, or MIBI, either alone or in combination, to correctly identify the preoperative location of PC is approximately 80% and reached 95% or more when the 3 procedures were used together[11] (Christakis I et al).

The single most effective therapy for parathyroid carcinoma is complete resection of the primary lesion at the time of initial operation when extensive local invasion and distant metastases are less likely. When the gross pathological findings suggest such as PC has a white, rough surface attached to the nearby tissues and is usually bigger than Parathyroid Adenoma. For malignancy, en bloc removal of the lesion together with the ipsilateral thyroid lobe and isthmus should be carried out[12](shane E et al.).

Although en bloc resection is the preferred method of treatment in patients with PC, as brown tumors are believed to be dependent on PTH, resection of parathyroid carcinoma generally marks spontaneous regression. Occasionally, local excision is only recommended when regression is incomplete or there is concomitant disfiguration of the affected bone[12].

Metastases occur via hematogenous spread and via the lymphatics. Lymph node metastasis occurs in ~15–30% of patients at their initial presentation. About 33% of patients have distant metastatic disease,

usually in the lungs, liver, and bone. The 5-year and 10-year overall survival rates are 82.3 and 66%, respectively. Median survival is 14.3 years with a median follow-up of 65.1 months (range 1 month–26 years)[13](Asare et al.).

4.CONCLUSION

Parathyroid Carcinoma presenting directly with the brown tumor in the maxilla (skeletal manifestations) without palpable neck mass or symptoms of hypercalcemia is extremely rare. Skeletal manifestations generally regress after the removal of parathyroid primary. Treatment of choice and bisphosphonates are essential for controlling the hypercalcemia and concomitant symptoms. Brown tumors in parathyroid carcinoma usually regress following parathyroid extirpation.

5.Conflict of interests

The authors declare that they have no competing/conflict of interests.

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