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Osteitis Fibrosa Cystica of Tibia as Initial Manifestation of Primary Hyperparathyroidism

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Abstract

Background: Primary hyperparathyroidism is disease of excessive secretion of parathyroid hormone. Osteitis fibrosa cystica or brown tumor, complication of primary hyperthyroidism is characterized by fibrotic cystic bony changes. Skeletal manifestations in the form of brown tumors are rare occuring in less than 2% of patients suffering from any form of hyperparathyroidism. *Case Report*: We report a 40-year-old woman with lytic lesions in tibia, diagnosed as osteitis fibrosa cystica secondary to parathyroid adenoma. *Conclusion*: Patients with primary hyperparathyroidism are usually asymptomatic and clinical presentation of the lesion in tibia is rarely a first sign of the disease.

Keywords: Neoplasms, Osteitis Fibrosa Cystica, Parathyroid Hormone, Primary Hyperparathyroidism, Tibia.

Introduction

Osteitis fibrosa cystica is a skeletal disease related to long standing end-stage hyperparathyroidism and is sometimes referred to as "brown tumor" because of its reddish appearance. However, it is not a true neoplasm but rather a reactive osteolytic lesion of bone and may mimic other diseases such as giant cell tumor, multiple bony metastasis or multiple myeloma [1]. Primary hyperparathyroidism is defined as hypercalcemia from the overproduction of parathyroid hormone (PTH) by one or more hyperfunctioning parathyroid glands [2]. It can be caused by a solitary adenoma in 80% of patients, parathyroid hyperplasia in 15%, multiple adenoma in 5% and parathyroid carcinoma in less than 5% of patients [1]. Here we report a patient with primary hyperparathyroidism caused by parathyroid adenoma with osteitis fibrosa cystica of tibia as an unusual first manifestation of the disease.

Case Report

A 40 year old female patient presented to the orthopaedic out-patient department complaining of pain and swelling in the middle part of right leg since last three months. Plain X-ray revealed a well-defined lytic lesion with multiple thin internal septations in the cortex of mid shaft of right tibia **[Fig.1]**. Possibility of osteoblastoma, aneurysmal bone cyst and non-ossifying fibroma were suggested. MRI showed an expansile multiloculated lytic lesion arising eccentrically from the cortex of antero-medial aspect of right tibia, possibly adamantinoma **[Fig.2,3]**. Fine needle aspiration cytology of the lesion showed features of giant cell lesion and further investigations with excision biopsy were suggested **[Fig.4]**. The biochemical investigations showed elevated serum calcium levels of 13.3 mg/dL (normal 8.5-10.5 mg/dL), serum phosphorus of 1.6 mg/dL (normal

2.7-4.5 mg/dL), serum alkaline phosphatase of 622 IU/L (normal 64-306 IU/L) and serum parathyroid hormone of 703.20 pg/mL (normal upto 11.5 pg/mL). Excision biopsy of the lesion showed diffuse proliferation of osteoclast-type giant cells, increased blood vessels and areas of haemorrhage **[Fig.5]**. Ultrasound scan of neck showed a well-defined hypoechoic lesion at inferior pole of right lobe of thyroid with increased vascularity. A possibility of right inferior parathyroid adenoma was suggested. MRI scan of neck confirmed the same findings with additional findings of multiple lytic lesions in the right clavicle, left scapula and right mandible. The Tc99m sestamibi scan showed increased uptake in right inferior parathyroid site **[Fig.6]**.

Based on the above findings the diagnosis of primary hyperparathyroidism with osteitis fibrosa cystica due to parathyroid adenoma was considered. Right inferior parathyroidectomy was done and the histopathological examination confirmed the lesion as parathyroid adenoma **[Fig.7]**. Two weeks postoperatively, the serum levels of calcium and parathyroid hormone declined to their normal limits.

Discussion

Osteitis fibrosa cystica also known as osteitis fibrosa, osteodystrophia fibrosa, Von Recklinghausen's disease of bone, is a skeletal disorder caused by prolonged exposure of bone to elevated parathyroid hormone levels in primary hyperparathyroidism [3]. Osteitis fibrosa cystica was first reported by von Recklinghausen in 1891; it presents as the end stage findings of hyperparathyroidism. However recently, with the technical development of imaging and laboratory screening methods, hypercalcemia due to primary or secondary hyperparathyroidism can often be detected early; as a result the frequency of osteitis fibrosa cystica has declined [1]. Lytic lesions caused by hyperparathyroidism are called brown tumors. The term "Brown tumor" is a misnomer because it is not a true neoplasm, but

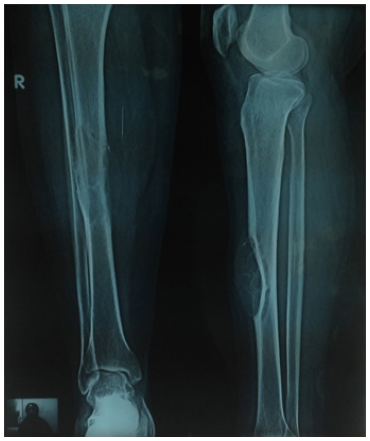

Fig.1: *Well defined expansile lytic lesion involving the anterior cortex of mid shaft of tibia.*

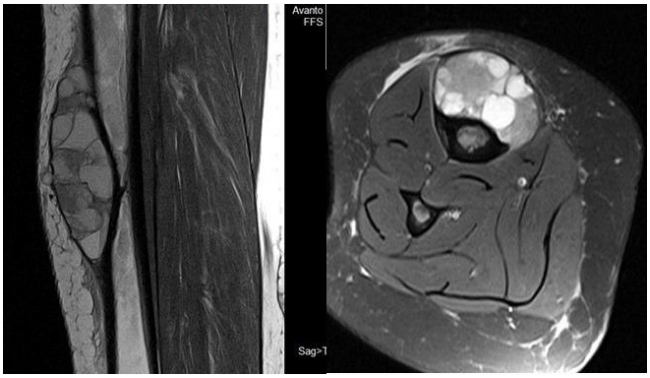

Fig.2: *T2 sagittal and T2 FS axial MRI showing multiseptated heterogenous signal intensity lesion arising from cortex of tibia.*

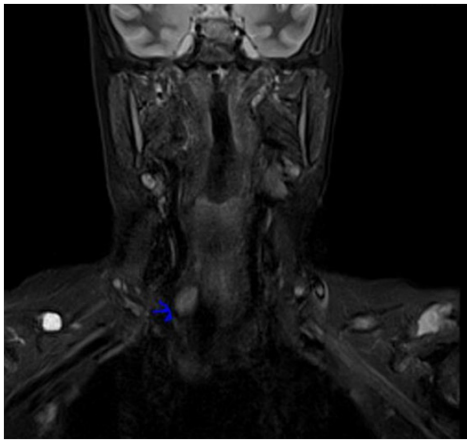

Fig.3: *T2FS coronal MRI of neck showing hyper-intense parathyroid adenoma at the lower pole right thyroid gland.*

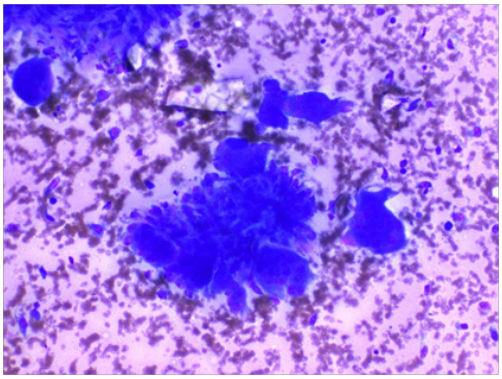

Fig.4: *FNA smear showing many multinucleated giant cells admixed with stromal cells in a background of haemorrhage. (Giemsa, 20X).*

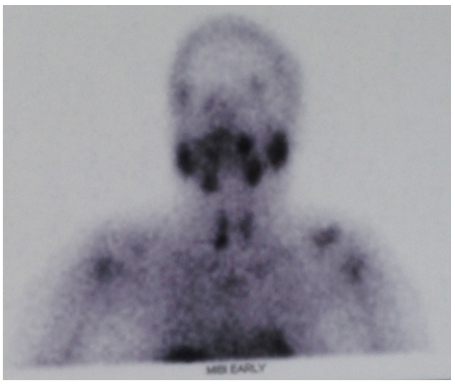

Fig.6: *MIBI image showing focal increased radiotracer uptake in relation to lower pole of right lobe of thyroid.*

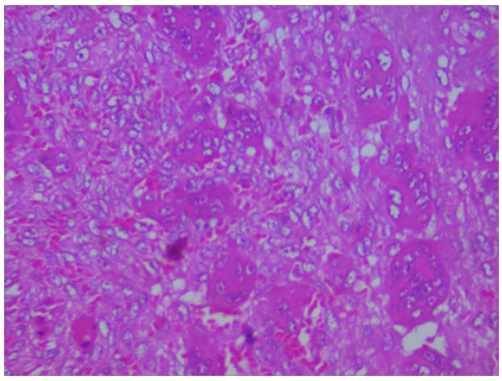

Fig.5: *Section shows diffuse proliferation of osteoclast-type giant cells admixed with spindle cell proliferation and haemorrhage (H&E, 20X).*

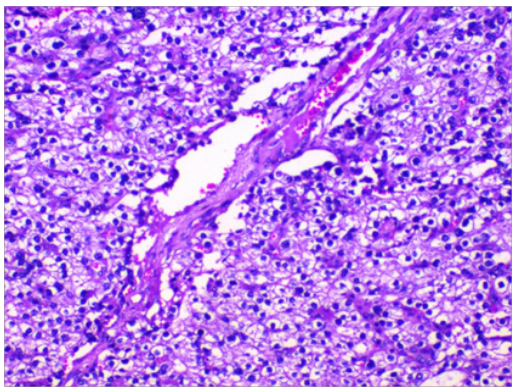

Fig.7: *Section shows diffuse sheets of round to polygonal cells having clear cytoplasm and uniform nuclei (H&E, 10X).*

benign hypervascular focal lesions in bones caused by increased osteoclastic activity and fibroblastic proliferation [4]. They are seen commonly in facial bones, mandible, sternum, pelvis, ribs, femur and rarely in vertebrae [3].

Primary hyperparathyroidism is characterized by the hyperfunctioning of parathyroid glands. The female:male ratio is 5:2. About 85% of cases are caused by a sporadic PTH secreting solitary adenoma of parathyroid chief cells. Multiglandular parathyroid hyperplasia occurs in 1-15% of patients with primary hyperparathyroidism. Parathyroid carcinoma is rare and occurs in less than 1% of cases. Although this condition can occur at any age, it commonly affects people over the age of 50 years and postmenopausal women [5,6]. Excessive secretion of PTH causes bone resorption accompanied by fibrovascular marrow replacement and increased osteoblastic activity; the imbalance of osteoclastic and osteoblastic activity manifest as a slow enlarging painful bony mass, osteitis fibrosa cystica, which can lead to pathological fractures. Fine needle aspiration biopsy (FNAB) of the mass-like lesion, imaging studies, high serum calcium and PTH leves are useful for diagnosis. Treatment of osteitis fibrosa cystica caused by primary hyperparathyroidism is surgical removal of the parathyroid lesions.

Conclusion

Osteitis fibrosa cystica has become a rare disease and lytic lesion of bone showing giant cells in histopathology should raise the suspicion of hyperparathyroidism. A high index of suspicion will lead to early diagnosis.

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