Case Report

Solitary phalangeal brown tumour in primary hyperparathyroidism: Report of a rare presentation

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ABSTRACT

Parathyroid adenoma is the most common cause of primary hyperparathyroidism. Severe cases of primary hyperparathyroidism manifest as osteitis fibrosa cystica generalisata, characterized by generalized bone loss with increased bone resorption, including both subperiosteal and endosteal surfaces. The most common sites for formation of fibrotic cystic lesions (brown tumors) are in the long bones and jaw which present as swelling, pathological fracture, and/or bone pain, usually involving multiple sites. Here, we describe an unusual presentation of a solitary brown tumor in a young male who initially presented to the hand surgeon with a history of right thumb swelling following trivial trauma. Further detailed clinical, biochemical, scintigraphic (Tc 99m methylene diphosphonate scintigraphy and Tc 99m Sestamibi scintigraphy), and radiological investigations aided definitive diagnosis and treatment. The causative parathyroid adenoma was excised curing hyperparathyroidism and the lesion regressed substantially.

Keywords: Brown tumor, parathyroid adenoma, Tc 99m methylene diphosphonate bone scintigraphy, Tc 99m sestamibi scintigraphy

INTRODUCTION

Primary hyperparathyroidism is a disorder which presents as a spectrum of signs and symptoms due to abnormality in calcium, phosphate, and bone metabolism caused by increased secretion of parathormone (PTH).[1] Increase in PTH results in hypercalcemia and hypophosphatemia. There can be variable presentations including recurrent nephrolithiasis (10-25%), peptic ulcers, mental changes, and less frequently, extensive bone resorption (10-20%), resulting in multiple fibrotic cystic lesions (brown tumors).[2] Here, we describe an unusual presentation with a solitary brown tumor in a young male with primary hyperparathyroidism.

CASE REPORT

A 39-year-old male was referred to a hand surgeon at our hospital with history of swelling in the right thumb of 2 years duration. It was noticed following a trivial trauma and it had been gradually increasing in size with no associated constitutional symptoms. On clinical examination, there was grade 4 clubbing of right thumb [Figure 1a]. A plain radiograph of the hand showed an expansile lytic lesion involving the distal phalanx of the thumb with marked cortical thinning and focal defects in the cortex. Subperiosteal bone resorption was seen in the phalanges, particularly along the radial aspects of the middle phalanges of the middle and index finger and also along the phalangeal tufts [Figure 1b]. In view of elevated serum alkaline phosphatase and with a suspicion of solitary bone metastasis, Tc 99m methylene diphosphonate

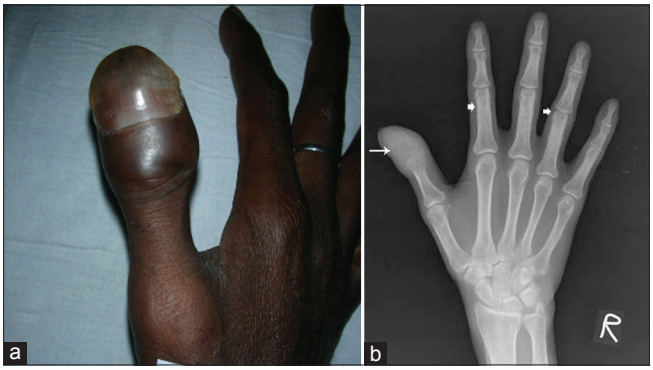

Figure 1: (a) Unilateral clubbing of left thumb Grade 4. (b) Anterior-posterior radiograph of the right hand showing a lytic expansile lesion involving the distal phalanx of the thumb with marked thinning deficiency in the cortex (arrow). Subperiosteal bone resorption is seen in the phalanges, particularly along the radial aspects of the middle phalanges of the middle and index finger (two short arrows) and also along the phalangeal tufts (arrow heads)

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bone scintigraphy was performed. Bone scintigraphy revealed generalized intense tracer uptake in the axial and appendicular skeleton including the costal cartilages [Figure 2a]. Increased tracer uptake was noted in the distal phalanx of right thumb [Figure 2b] and both scapulae. There was stasis of tracer in both kidneys. Based on these findings, the possibility of hyperparathyroidism was suggested. Further evaluation revealed elevated serum calcium, 12.5 g% and serum PTH levels, 1,821 pg/ml.

Radiograph of the lumbar spine showed diffuse osteopenia. Bilateral stag horn renal calculi were also noted [Figure 3a]. Lateral radiograph of the skull showed granular deossification giving the classical "salt and pepper" appearance and loss of sharp definition of the inner and outer tables [Figure 3b]. Tc 99m sestamibi parathyroid scintigraphy [Figure 4a] demonstrated a functioning parathyroid adenoma in the region of the inferior pole of left lobe of thyroid, which was confirmed by single-photon emission computed tomography [Figure 5a and b] with associated tracer uptake in the distal phalanx of the right thumb [Figure 4b] confirming the diagnosis of primary hyperparathyroidism due to left inferior parathyroid adenoma and brown tumor right thumb [Figure 6]. Ultrasound examination of the neck revealed a 10 cm × 6 mm sized well-defined ovoid hypoechoic lesion at the level of the lower pole of left lobe of thyroid, but lateral to the carotid artery, between the common carotid artery (CCA) and internal jugular vein (IJV). There was marked vascularity within the lesion on Doppler image [Figure 6]. This corresponded to the lesion seen on the Tc 99m Sestamibi scan and an ectopic location of parathyroid adenoma was considered.

He underwent excision of left inferior parathyroid adenoma. The operative findings revealed 2 cm × 1 cm lobulated left inferior parathyroid adenoma in ectopic location within left carotid sheath between the carotid artery and IJV butting the vagus nerve and also a 1.5 cm whitish firm nodule superior to it which was thought likely to be ectopic thymic remnant. The findings were confirmed by histopathology. His post-operative corrected serum calcium level was within normal limits (8.3 mg%). His follow-up after 1 year revealed 50% reduction in the size of the thumb swelling and his follow-up serum calcium was 9.4 mg%.

DISCUSSION

Metabolic bone diseases refer to a broad spectrum of diseases caused by abnormalities of minerals such as calcium, phosphorus, magnesium, or vitamin D leading to variety of disorders. Clinical conditions most commonly referred to as metabolic bone disorders are osteoporosis, Paget's disease of bone, osteomalacia (adults) and rickets (children), Fibrous dysplasia, and osteitis fibrosa cystica. The bone disease osteitis fibrosa cystica was first described by von

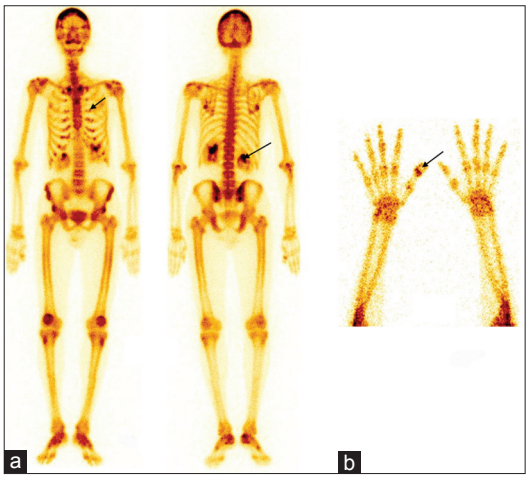

Figure 2: (a) Tc 99m methylene diphosphonate (MDP)-whole body bone scintigraphy showing diffuse increase in tracer uptake in the skull and long bones of both upper and lower limbs and in costal cartilages. (b) Spot view Tc 99m MDP bone scintigraphy of both hands showing photopenic area with surrounding increased tracer uptake in the distal phalanx of right thumb

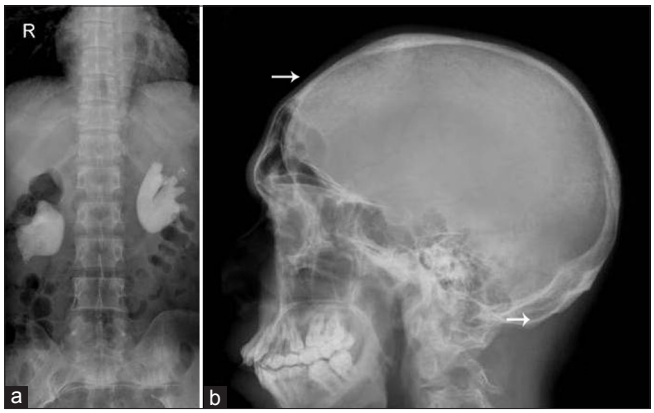

Figure 3: (a) Anterior-posterior radiograph of the lumbar spine showing, bilateral staghorn calculi (arrows). Osteopenia in the vertebrae is also noted. (b) Lateral radiograph of the skull showing granular deossification giving the classical æsalt and pepperæ appearance (arrows) and loss of sharp definition of the inner and outer tables

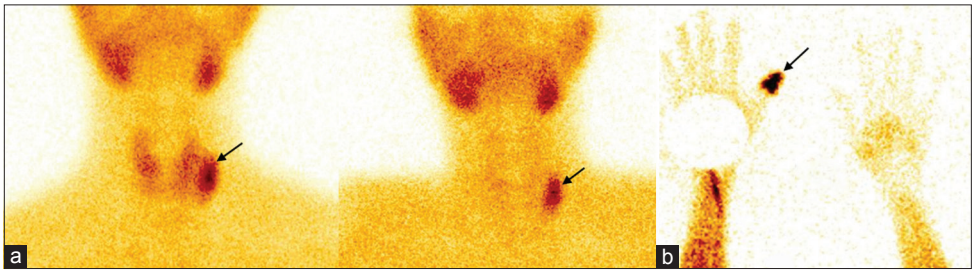

Figure 4: (a) Tc 99m MIBI whole body scan showing left inferior parathyroid adenoma. (b) Spot view Tc 99m MIBI scintigraphy of both hands showing increased tracer uptake in the distal phalanx of right thumb

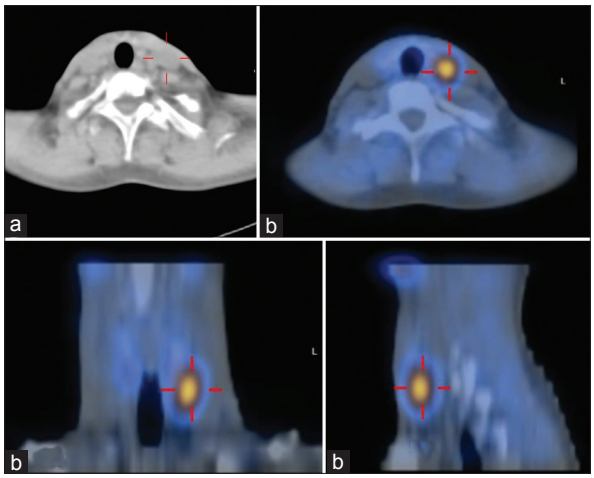

Figure 5: (a- b) Single-photon emission computed tomography images confirming the exact anatomical location of the parathyroid adenoma

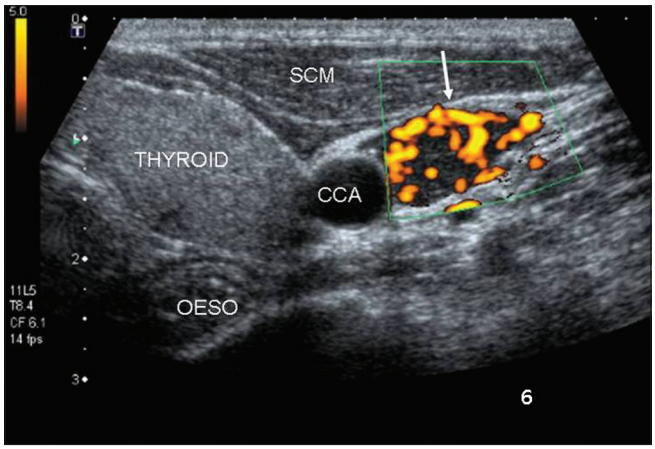

Figure 6: Transverse ultrasound Doppler image of the neck at the level of lower pole of thyroid, showing a well-defined hypo echoic lesion posterior lateral to the carotid artery (relationships as shown). Power Doppler shows marked vascularity within the lesion

Recklinghausen in 1891, but the etiological link between the disease and parathyroid neoplasms was not established until 1925.[1] Brown tumors are skeletal manifestations of hyperparathyroidism that may mimic metastasis. They are clinical consequence of untreated severe primary or secondary hyperparathyroidism. Brown tumors are composed of numerous multinucleated osteoclasts (giant cells) admixed with stromal cells and matrix, which are found most often in trabecular portions of the jaw, long bones, and ribs. Radiographic features described in hyperparathyroidism are mainly due to bone resorption, which could be subperiosteal, subchondral, subligamentous, cortical, and trabecular. Subperiosteal bone resorption, which gives a lace-like irregularity to the cortical margin, is classically described in the radial aspect of the middle phalanx of the 2nd and 3rd fingers, phalangeal tufts, neck of humerus and femur, and distal end of clavicle. Subchondral bone resorption results in pseudo-widening of the joint space and can be seen commonly in small joints of the hand, sacroiliac joints, acromioclavicular joints, and pubic symphysis. Cortical bone resorption gives rise to the appearance of intracortical tunneling and scalloping along the endosteal margins. Trabecular bone resorption results in spotty deossification and indistinct trabecular pattern. In the skull, this is seen as the granular "salt and pepper" appearance. Subligamentous bone resorption is seen at insertions of ligaments and tendons. Common sites of occurrence are in the pelvic bones, ribs, clavicles, and bones of extremities.[2] Bone scintigraphy is a highly sensitive method for the detection of the altered local bone metabolism; however, it lacks specificity.[3]

Benign disorders should be considered in patients presenting with solitary bone lesions and in whom there is no known malignancy. Isolated skeletal metastasis of unknown origin is a relatively rare entity.[4-6] Few unusual sites of brown tumor have been reported earlier.[7-10] and brown tumor should be included in the differential for locally increased bone metabolism. Correlation with the radiographic findings and biochemical parameters is helpful.

99m Tc sestamibi or 99-m Tc methoxyisobutylisonitrile MIBI scintigraphy is the radionuclide study of choice for pre-operative localization parathyroid adenoma. Because of superior image quality, more favorable dosimetry, and improved accuracy, 99m Tc sestamibi is the radio isotope most commonly used. High-quality scintigraphy with 99m Tc-sestamibi can accurately localize parathyroid adenomas in 85-95% of patient 11.[11] The retention of this tracer in parathyroid lesions is due to the presence of oxyphil cells in these lesions. These oxyphil cells are rich in mitochondria, which are the sites of intracellular MIBI sequestration. The "single isotope, double-phase technique" is the most commonly used technique which is based on differential washout of MIBI from thyroid and parathyroid glands. Additional single-photon emission computed tomography to planar scintigraphy provides information for localizing parathyroid lesions, to differentiate thyroid from parathyroid lesions, and especially in detecting and localizing ectopic parathyroid lesions 12.[12]

In ultrasound gray-scale images, parathyroid adenomas appear as discrete, oval, anechoic, or hypoechoic masses located posterior to the thyroid gland; anterior to the longus colli muscles, and frequently medial to the CCA 13.[13] An echogenic line that separates the thyroid gland from the enlarged parathyroid gland can usually be seen 14.[14] Color Doppler imaging showed marked vascularity within the adenoma, especially when the lesions are more than 1 cm in size.

The sensitivity of neck ultrasonography (USG) in identification of parathyroid adenomas ranges between 70% and 80% and sometimes may be as low as 40% in patients with prior surgical procedure. However, USG is useful to confirm the presence of a scan-positive solitary parathyroid adenoma.[15]

Other findings secondary to the hypercalcemia include medullary nephrocalcinosis or renal calculi, pancreatic calcifications, and calculi due to pancreatitis and metastatic calcifications.

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