

Severe Hyperparathyroidism with Secondary Osteitis Fibrosa Cystica and Brown Tumors Mimicking Bone Metastasis

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Abstract

Introduction: The classical presentation of primary hyperparathyroidism, osteitis fibrosa cystica, has become very rare nowadays. This rarity makes it sometimes misdiagnosed leading to disastrous outcomes.

Methods: We present a case of an undiagnosed primary hyperparathyroidism with severe osteitis fibrosa cystica and brown tumors that was first misdiagnosed as having bone metastasis.

Conclusion: Osteitis fibrosa cystica although rare should be considered in the differential diagnosis of patients presenting with multiple brown tumors, especially since the diagnosis can be easily made by a simple calcium level, thereby avoiding severe adverse outcomes.

Keywords: Primary hyperparathyroidism; Osteitis fibrosa cystica; Brown tumors; Hypercalcemia; Bone metastasis

Case Presentation

We present a case of a 31 year old female patient who was referred to our hospital for a swelling on her right lower leg (Figure 1).

History dates back to five years prior to presentation when the patient started having diffuse bony aches. Symptoms then started increasing in intensity until they started significantly affecting her mobility and quality of life for which she sought medical advice. Based on a Dual-energy X-ray absorptiometry scan she was informed of having severe osteoporosis for which intravenous zoledronic acid was started despite no work-up. She was kept on intravenous zoledronic acid for almost three years with no significant improvement in pain, moreover she started noticing an increasing swelling in her right lower extremity. Commuted Tomography Scan of the spine and pelvis were done and showed diffuse bony lesions in favor of a possible malignant process with diffuse bone metastases (images not available). Nevertheless patient refused any further medical treatment until she became bedridden and then was admitted by an oncologist to our institution for malignancy work-up. Her initial laboratory work-up revealed: Calcium 11.4; repeated: 11.2; Phosphorus 1.49; Albumin: 42; Magnesium: 1.46; creatinine: 0.39; spot calcium in urine: 20.2; spot creatinine in urine: 70.6; alkaline phosphatase 543; PTH 879.

Plain radiography of the right lower leg showed a multi-lobulated cystic bone lesion (Figure 2). MRI of the right lower leg showed a 6.6 × 4 × 3.3 cm multi-lobulated cystic mass with adjacent periosteal elevation occupying almost the whole part of the distal Right tibial shaft, consistent with a brown tumor (Figure 3). CT abdomen-pelvis showed multiple cystic bone lesions (iliac, sacral, femoral, ribs) also consistent with brown tumors (Figure 4).



Figure 1: The right lower leg mass (red circle).

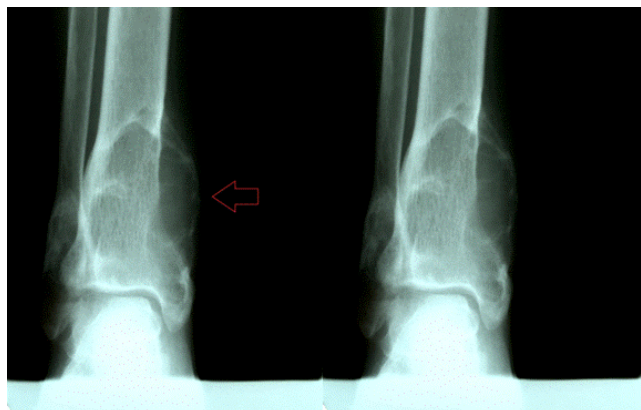


Figure 2: Right lower extremity X-ray showing the multilobulated cystic bone lesion (red arrow).

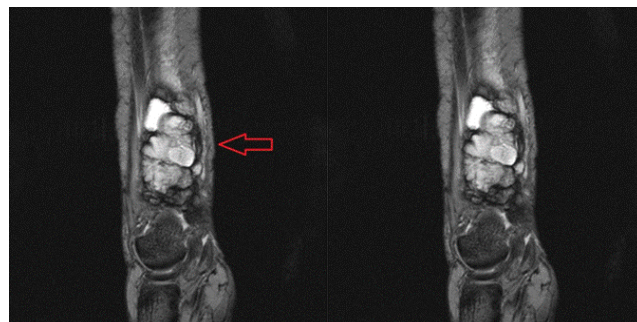


Figure 3: T2 right lower extremity MRI: 6.6 × 4 × 3.3 cm multilobulated cystic mass with adjacent periosteal elevation occupying almost the whole part of the distal Right tibial shaft, consistent with a brown tumor (red arrow).

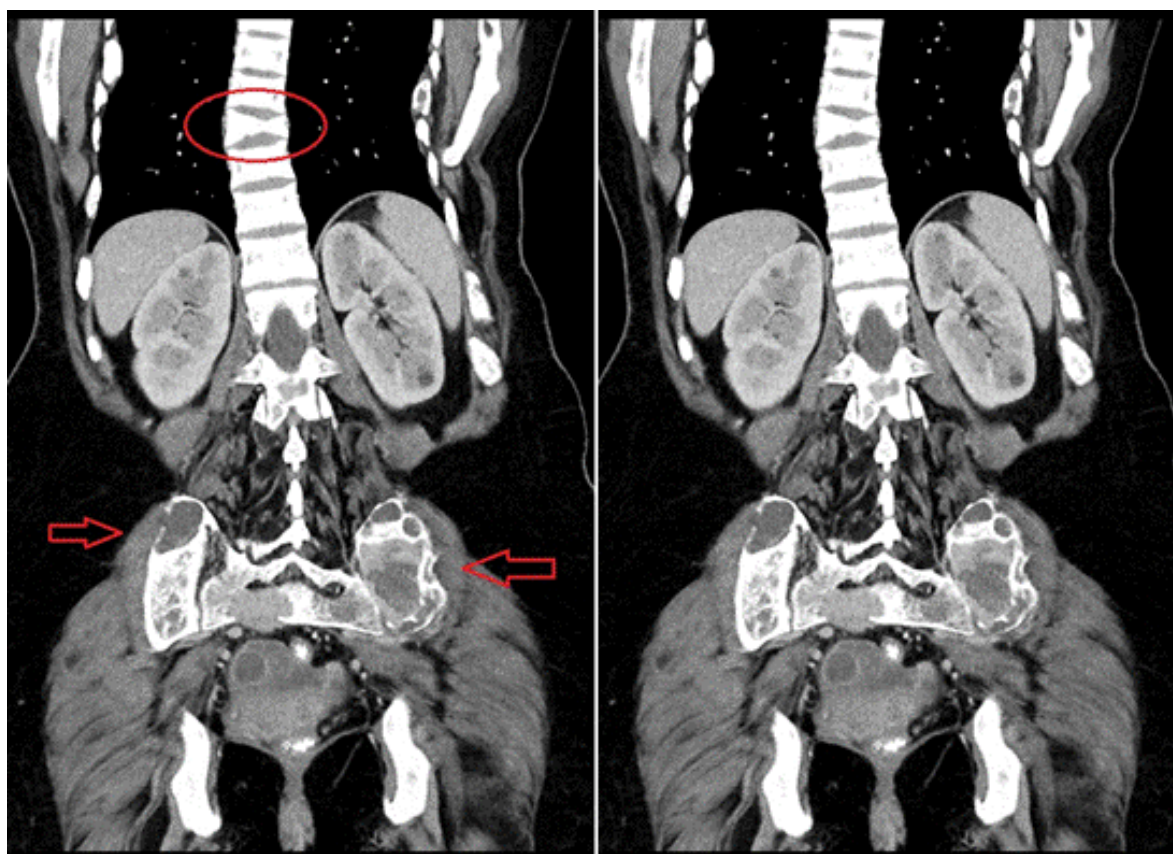


Figure 4: CT abdomen-pelvis showing a compression vertebral fracture (red circle) and several brown tumors (red arrows).

Patient was diagnosed with primary hyperparathyroidism with osteitis fibrosa cystica along with diffuse brown tumors. Parathyroid Tc99m-sestamibi scan revealed a focus of uptake on the lower pole of the left thyroid lobe evoking a hyper-functioning parathyroid gland (Figure 5). Patient then underwent parathyroidectomy with excision of the left lower parathyroid adenoma confirmed by pathology. Her

postoperative course was unremarkable except for a mild transient hypocalcemia secondary to hungry bone syndrome that responded well to treatment. A follow-up after one month of surgery showed major improvement in mobility and pain.

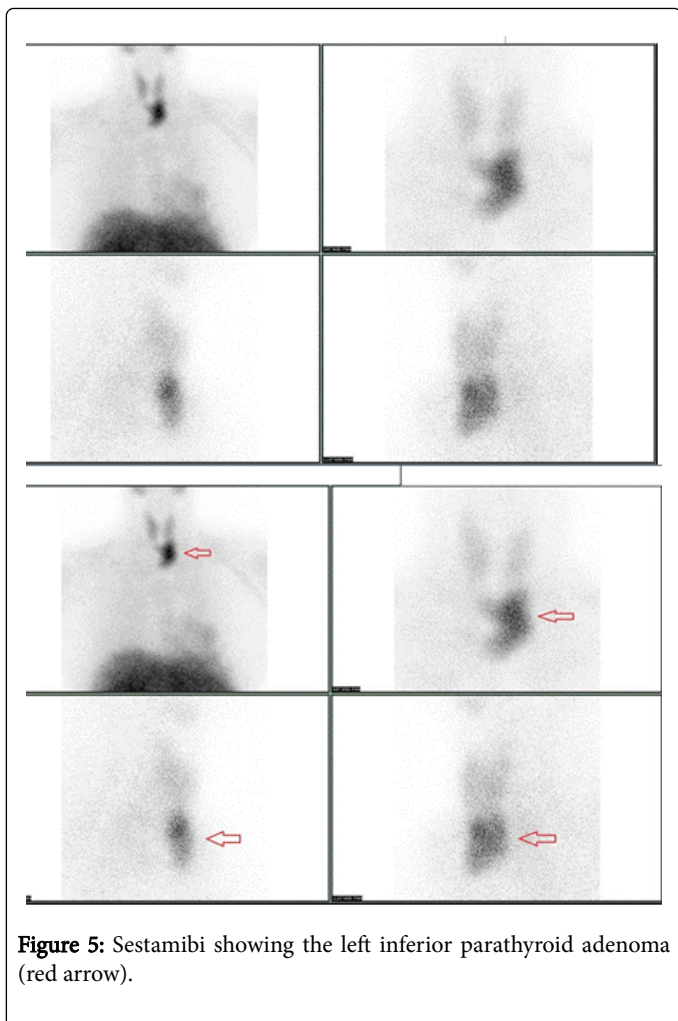


Figure 5: Sestamibi showing the left inferior parathyroid adenoma (red arrow).

Discussion

This is a case of severe undiagnosed primary hyperparathyroidism (PHPT) with osteitis fibrosa cystica mimicking bone metastasis.

With the increasing routine laboratory testing, the most common presentation of primary hyperparathyroidism nowadays is incidentally discovered hypercalcemia. When symptomatic, the prominent presenting features are usually non-specific and known as: “bone, stones, abdominal moans and psychic groans”.

Osteitis fibrosa cystica which is the classical presentation of primary hyperparathyroidism is extremely rare in this day and age especially in the United States and Europe. It occurs mostly in patients with severe

disease and is more commonly seen with parathyroid carcinoma [1]. In a review of 97 patients with mild PHPT, only one patient was found to have bone disease on routine conventional radiography [2]. However, in some Asian countries, unfortunately this entity is still commonly reported [3].

It is typically characterized by “salt-and-pepper” appearance in the skull, bone erosions and bone resorption of the phalanges, brown tumors and cysts of the long bones [4]. Brown tumors result from excess osteoclast activity and consists of collections of osteoclasts intermixed with fibrous tissue and poorly mineralized woven bone [5]. Histologically they are similar to giant cell tumors: multinucleated giant cells in a background of spindle cell proliferation containing a large amount of hemosiderin [6].

If undiagnosed this entity can lead to severe osteoporosis, diffuse pathological fractures, severe bone aches that can result in restriction of mobility ultimately ending in complete bed-rest similar to our described patient.

The challenge in the above described case at the time of presentation was the rarity of osteitis fibrosa cystica that led to misdiagnosis and mismanagement ultimately resulting in severe adverse outcomes to our patient.

Conclusion

Despite the fact that osteitis fibrosa cystica and brown tumors are rare manifestations of PHPT, they should be always kept in the differential diagnosis of bone lesions, especially since they can be easily diagnosed by a simple calcium level and if left untreated can lead to severe bone disease, deformities and fracture.

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