Investigating Unexpected Shortening of Height of A Middle-Aged Woman: A Case Report

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ABSTRACT

Osteitis fibrosa cystica (OFC) is a late sequela of untreated hyperparathyroidism caused by the autonomous secretion of parathyroid hormone (PTH). It is characterized by bone pain, skeletal deformities, and pathological fractures. OFC is caused by hypersecretion of PTH, leading to demineralization of bone and pathological fractures. We report a case of unusual presentation of OFC in a middle aged woman who noticed gradual shortening of her height within a particular period of time. Apart from height reduction, the patient had weight loss and pain in different parts of the body for over a decade. Despite being treated with analgesics and steroids initially, there was no significant improvement in her condition. Treatment history included surgical reduction of right elbow fracture and empirical anti-tubercular drug twice as there was a suspicion of disseminated tuberculosis. The patient developed lytic changes, skeletal deformities, and osteoporosis, and was diagnosed with a solitary parathyroid adenoma on the right side.

Keywords: Parathyroid hormone, skeletal deformity, Osteitis fibrosa cystica, pathological fractures, autonomous secretion, parathyroid adenoma.

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INTRODUCTION

Osteitis fibrosa cystica (OFC) is a late complication of untreated hyperparathyroidism caused by a solitary parathyroid adenoma secreting parathyroid hormone (PTH) autonomously. OFC is characterized by bone pain, skeletal deformities, and pathological fractures. Other common bone diseases, such as primary bone malignancies and tuberculosis, sometimes manifest the same symptoms as OFC. The pathophysiology of OFC includes hypersecretion of PTH, which leads to the release of calcium from bone cells by inhibiting osteoblasts and stimulating osteoclast activities, causing demineralization of bone and consequently making it prone to pathological fractures (1). Secondary osteoporosis is the result of an underlying disease, such as an endocrine abnormality, neoplasm, or drugs that adversely affect bones. One of these endocrine abnormalities is primary hyperparathyroidism. The most common cause of this abnormality is an adenoma of the parathyroid gland (2).

CASE REPORT

A 38-year-old woman noticed gradual shortening of her height over a period of 10 years, which was associated with weight loss and polyarthralgia for approximately same period. Non-diabetic, normotensive homemaker and mother of two healthy children stated that she was 5 feet 2 inches at the time of her wedding in 2004 but 4 feet 6 inches at present (Figure 1).

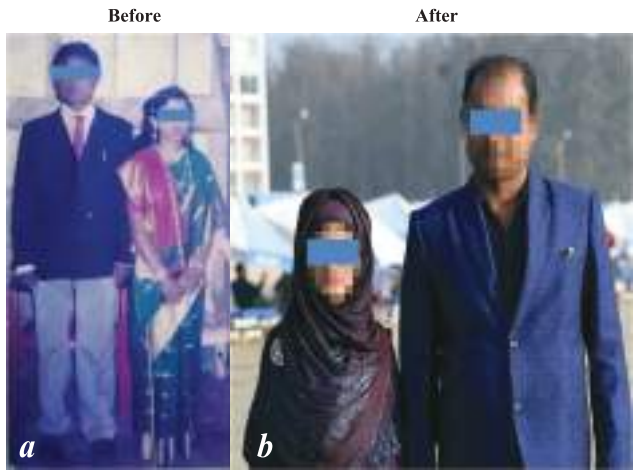


Figure-1: a) Past image (2004) of the patient with her husband. Reached up to the earlobe of husband. b) Present image (2022) showing that she reaches up to the shoulder level of her husband. (Image shared with informed consent)

She mentioned experiencing pain that started in her right hip and right thigh, later radiated to the back and right leg. Initially diagnosed with polyarthritis, she was treated with analgesics and steroids for a short period of time. It should be noted that, at the same time, she reported losing 24 kg over the last 12 years. For the first 4 years, her weight loss was accompanied by intermittent fever, which was not associated with chills and rigors and remitted spontaneously. An empirical anti-tubercular drug regimen was prescribed to the patient twice: once in 2016 with 2HRZE 4HR and again in 2020 with 1HRZE 5HRE. Although there was a short-term improvement in her general physical condition, it did not last long. Her past medical history is significant for a low-trauma fracture of the right elbow in 2014, which was fixed by open reduction and internal fixation. Her general examination revealed a squint, left forearm bowing (Figure 2 b), and a surgical scar in her right elbow.

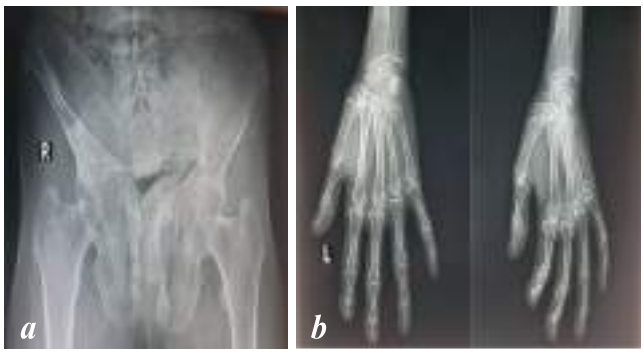


Figure-2: X-ray images of a) pelvis and b) hands-showing lytic changes & acro-osteolysis of the distal phalanges suggesting generalized osteopenia. Bowing of left forearm is seen.

The following anthropometric measurements were provided by the patient: weight = 31.5 kg, height = 4 feet, 6 inches (138 cm) with a BMI of 16.7 kg/m^2. Systemic examinations were unremarkable. Locomotor system examination (GALS scoring) revealed limpic gait, regression of nails in fingers with shortening and tufting of distal phalanges, restricted hip movements, forward indentation of the vertebral column in the lumber region with scoliosis, and a normal Schober's test. A set of laboratory investigations was done, which illustrated increased levels of serum calcium, PTH, and alkaline phosphatase and decreased levels of inorganic phosphate and vitamin D. An X-ray of the pelvis showed deformity and dysplasia, reduced joint space, and generalized osteopenia, features that are consistent with OFC. A bone mineral density test revealed T-scores of -6.1, -3.8, and -3.6 in the radius, spine, and femur, respectively, indicative of osteoporosis.

A high-resolution neck ultrasound revealed a small (measuring about 22 X 12 X 10mm) but well outlined, nearly ovoid, solid, hypoechoic soft tissue lesion located posterolateral to the lower pole of the right thyroid lobe (Figure 3).

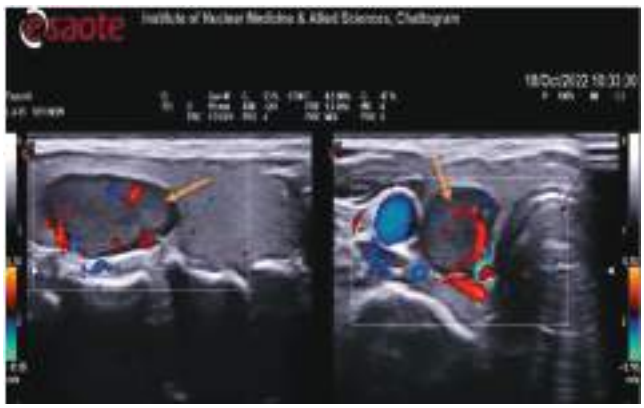
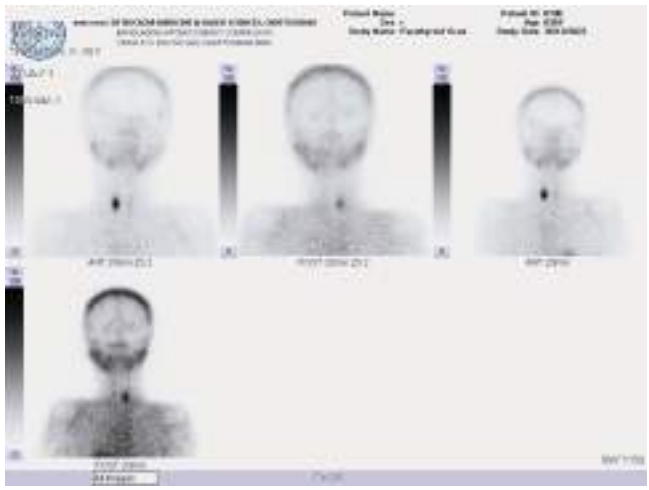


Figure-3: High resolution neck ultrasound image showing a well-outlined, nearly ovoid, hypoechoic and hyper-vascular lesion (arrow), postero-lateral to right thyroid lobe, with low resistance arterial flow in spectral Doppler

Subsequently, the ^{99m}Tc-sestamibi parathyroid scan showed a focal area of increased radiotracer concentration in the region of the lower pole of the right lobe of the thyroid gland in the early planar image at 20 minutes. A delayed image showed persistent tracer activity in the above-mentioned site with complete washout from the rest of the area at 120 minutes (Figure 4); thus, a solitary parathyroid adenoma was diagnosed, but SPECT-CT and CT imaging correlation were also done (Figure 5).



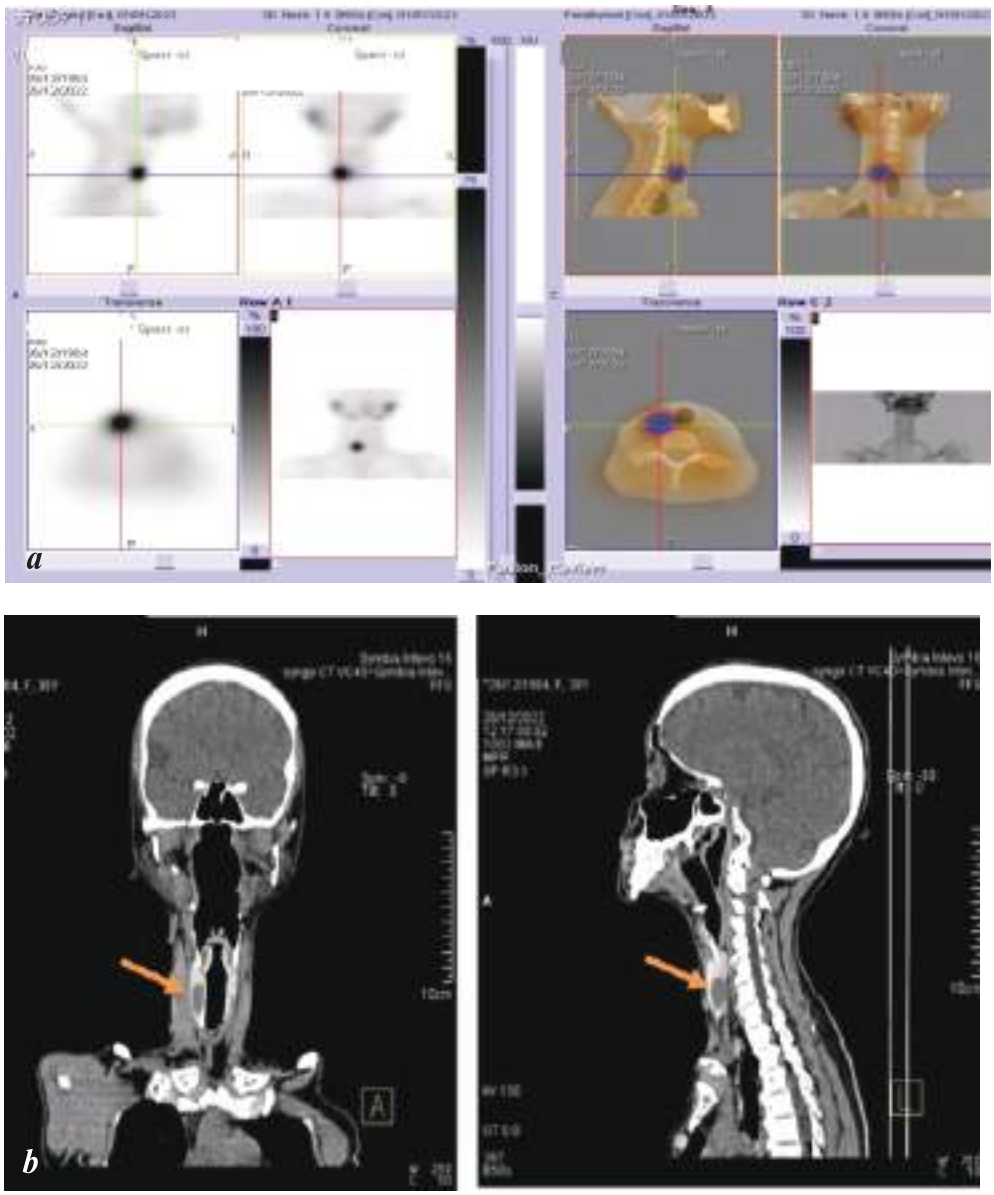


Figure 5: a) Delayed (120 minutes) images of ^{99m}Tc-sestamibi parathyroid scintigraphy in the same patient showed persistence of tracer uptake in the right lower pole area of the thyroid gland. b) Corresponding CT images of parathyroid adenoma (arrow) in coronal and sagittal planes demonstrating the precise location of an ovoid mass in the lower right parathyroid region.

DISCUSSION

Osteitis fibrosa cystica (OFC) is a rare osteolytic and histologically benign metabolic bone disease that occurs in approximately 5% of the patients with primary hyperparathyroidism (3). This condition results from increased bone resorption by osteoclasts with fibrous replacement in the lacunae and is characterized by bone pain and tenderness, fractures, and deformity (4). The activation of osteoclasts leads to trabecular remodeling and loss of cortical bone, leading to osteoporosis, which is the most common skeletal manifestation of hyperparathyroidism. Clinical features that contributed to the diagnosis of primary hyperparathyroidism in this reported case were musculoskeletal impairment, generalized osteoporosis (confirmed by x-ray and bone mineral density examination), increased PTH, and hypercalcemia. A high-resolution ultrasound examination found a definite nodular lesion in the right parathyroid gland. Parathyroid scintigraphy also concluded that there

was an area of increased tracer uptake in the lower right parathyroid gland region that was suspected as an adenoma. CT correlation was also done to precisely locate the adenomatous lesion. From the literature, it is said that the most common lesion found in patients with primary hyperparathyroidism is a solitary parathyroid adenoma, occurring in 80% of patients (5).

Primary hyperparathyroidism is due to increased blood concentrations of endogenous parathyroid hormone (PTH), secondary to a benign parathyroid adenoma (6). Skeletal X-rays remain mostly unremarkable in milder primary hyperparathyroidism; however, in advanced disease, characteristic changes such as demineralization, subperiosteal erosion, and terminal resorption in the phalanges can occur. Clinically, OFC can also be associated with pathological fractures (7). Bone involvement in hyperparathyroidism has shown a significant decrease in incidence over the past decades (from 80% to as low as 15%). This fact might be due to the early detection of asymptomatic cases via serum calcium monitoring (8) Osteitis fibrosa cystica is frequently misdiagnosed as other bone lesions, particularly malignant tumors such as osteosarcoma, Ewing sarcoma, lymphoma, multiple myeloma, metastatic carcinoma, and disseminated tuberculosis, and should be considered in the differential diagnosis. (9) Although diagnosing OFC can be difficult, if treated properly, this condition is reversible (10). The symptoms of primary hyperparathyroidism disappear if the adenoma of parathyroid gland is removed surgically. Since this is the only definitive approach to primary hyperparathyroidisism, surgery is the standard treatment of this disease. The primary treatment for a parathyroid adenoma is parathyroidectomy as the symptoms are due to hyperparathyroidism. Surgical excision is the primary and only form of treatment once the results of the biochemical and radiographic study reveal the presence of a parathyroid adenoma. PTH levels returning to normal within 10 minutes of surgery is conclusive evidence of successful parathyroidectomy /excision of the parathyroid adenoma.

CONCLUSION

The insight of this case lies in the fact that it illuminates the necessity of assessing parathyroid function in patients with bone lesions.

In the rare 5% of cases where primary hyperparathyroidism is symptomatic at the skeletal level, it usually presents as OFC. Elevated PTH and hypercalcemia complement the radiological and histological findings to establish the diagnosis of OFC.

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