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A case of parathyroid carcinoma: the rarest endocrine cancer

Chowdhury C[1], Das S[2], Kar A[3], Kashem CT[4], Uddin MR[5], Mallick S[6], *Akter F[7]

[1]Chirantan Chowdhury, Post Graduate Trainee, Department of Endocrinology, Chittagong Medical College and Hospital, Chattogram, Bangladesh; [2]Shemee Das, Post Graduate Trainee, Department of Endocrinology, Chittagong Medical College and Hospital, Chattogram, Bangladesh; [3]Antara Kar, Post Graduate Trainee, Department of Endocrinology, Chittagong Medical College and Hospital, Chattogram, Bangladesh; [4]Chowdhury Tanvir Ibne Kashem, Assistant Register, Department of Endocrinology, Chittagong Medical College and Hospital, Chattogram, Bangladesh; [5]Md. Rafiq Uddin, Assistant Professor, Department of Endocrinology, Chittagong Medical College and Hospital, Chattogram, Bangladesh; [6]Satyajit Mallick, Assistant Professor, Department of Endocrinology, Chittagong Medical College and Hospital, Chattogram, Bangladesh; [7]Farhana Akter, Associate Professor and Head of the Department of Endocrinology, Chittagong Medical College and Hospital, Chatttogram, Bangladesh

Abstract

Parathyroid neoplasm is the rarest endocrine cancer and an infrequent cause of primary hyperparathyroidism, responsible for less than 1% of cases. A 32-year-old male presented with neck swelling, generalized body aches, pain in multiple joints, and weight loss. When USG found a solid nodule in the left lobe, he underwent left hemithyroidectomy with excision of the left superior parathyroid mass but parathyroid and calcium level was persistently elevated and a whole-body scan suggested metastatic bone diseases. Finally, the completion thyroidectomy and excision of the remaining parathyroid mass were done successfully. One month after surgery patient became hypothyroid while parathyroid hormone and calcium came back to normal. He was diagnosed as a case of primary hyperparathyroidism due to parathyroid carcinoma with bone metastasis. *[J Assoc Clin Endocrinol Diabetol Bangladesh, July 2022; 1 (2): 65-68]*

Keywords: Parathyroid neoplasm; Hyperparathyroidism

***Correspondence:** Farhana Akter, Associate Professor and Head of the Department of Endocrinology, Chittagong Medical College and Hospital, Chatttogram, Bangladesh. Email: fakter36@gmail.com, Cell no: +8801711397888

Introduction

Primary hyperparathyroidism is part of a spectrum of parathyroid proliferative disorders that includes parathyroid adenoma, parathyroid hyperplasia, and parathyroid carcinoma. About 80-85% of cases account for parathyroid adenoma, 15% for hyperplasia, and 0.5-1% for carcinoma.[1] Parathyroid carcinoma is an infrequent neoplasm, with a prevalence of 0.005% of all cancers.[3] Most patients affected with this rare malignant tumor present with either sporadic primary hyperparathyroidism or primary hyperparathyroidism in the context of a genetic endocrine syndrome. Diagnosis is generally confirmed only after surgery by detailed pathological analysis unless there is preoperative evidence of gross local invasion, cervical lymph nodes, or distant metastases. The surgical approach offers the best disease control rates.[2,4-6] The most serious potential complications of parathyroid surgery. i.e. vocal cord paralysis and permanent hypoparathyroidism occur in fewer than 1% and 4% of procedures respectively when performed by highly skilled surgeons. Intravenous bisphosphonates have been used successfully to prevent post-operative complications.[7]

Case presentation

A 32-year-old male presented with generalized body aches associated with deep-seated bony pain in multiple joints for the last 5 months. There was no bone deformity. The patient had also complained of tiredness and loss of appetite causing a significant weight loss of around 18 kg within this period. He had noticed swelling in the front and left side of the neck for the last 1 month, which was initially small and painless but gradually increased in size over the last 15 days. He had no history of diabetes mellitus, hypertension, or dyslipidemia. There were no complaints of dysphagia, hoarseness, or palpitation.

With these complaints when the patient got admitted to the hospital, a local examination was done which

Table-I: Laboratory findings of the patient

Investigations	Initial evaluation	1st follow-up	2nd follow-up	1-month after 2nd surgery	Reference values
TSH (µIU/L)	1.638	-	-	53.89	0.5-5.0
FT3 (pmol/l)	3.76	-	-	-	4.6-9.7
FT4 (pmol/l)	11.86	-	-	0.35	10.0-28.0
S. Calcium (mg/dl)	14.80	9.97	9.92	8.64	8.5-10.5
S. PTH (pg/ml)	943	516	368	59	10.0-55.0
S. Phosphorus (mmol/l)	1.2	-	-	-	1.12-1.45
S. CPK (U/L)	118	-	-	-	39.0 – 308.0
Creatinine (mg/dl)	1.3	-	-	-	0.74-1.35
Uric Acid (mg/dl)	7.0	-	-	-	3.5-7.2
RA factor	Negative	-	-	-	-
HLA-B27	Negative	-	-	-	-
25-OH Vit-D (ng/ml)	-	-	23.53	-	20.0-40.0
S. Albumin (g/dl)	-	-	-	4.38	3.4-5.4

TSH: thyroid stimulating hormone, PTH: parathyroid hormone, CPK: creatine phosphokinase

revealed there was a solitary swelling in the thyroid region, on the left side of the neck about 2×2 cm in size, globular in shape, smooth surface, firm in consistency, mobile from side to side and above downwards, the margin was well defined, the temperature was normal, non-tender, overlying skin & underlying structure was free, the trachea was central in position, get below the swelling was possible, no audible bruit. On general examination during admission: pulse: 76 /min, blood pressure: 110/70 mm of Hg, temperature: 98.4ºF, respiration: 14 breaths/min. There were no palpable lymph nodes. His initial lab investigations are shown in Table-I.

An Ultrasonogram (USG) of the thyroid gland revealed a fairly large lobulated solid nodule (41×24 mm) at the upper pole of the left lobe (Figure 1). Fine needle aspiration cytology (FNAC) of the left lobe of the thyroid gland showed thyroid follicular cells in clusters and singly mixed with inflammatory cells and macrophages. Some of the follicular cells show hurtle cell changes. Nuclear crowding and overlapping were present. Parathyroid scintigraphy was negative for parathyroid adenoma or hyperplasia (Figure 2). USG of the whole abdomen revealed calcification at both renal pyramids (Figure 3). A skeletal survey of this patient revealed granular 'salt and paper' appearance in the skull, multiple bony osteolytic expansile lesions with cortical thinning, diffuse osteoporosis without any pathological fracture in the following bones: head of 5th metacarpal (left), head of proximal phalanges of the middle finger (left), body of the pubis, base and adjacent shaft of 5th metatarsal (right).

After the evaluation, the patient underwent left hemithyroidectomy with excision of the left superior

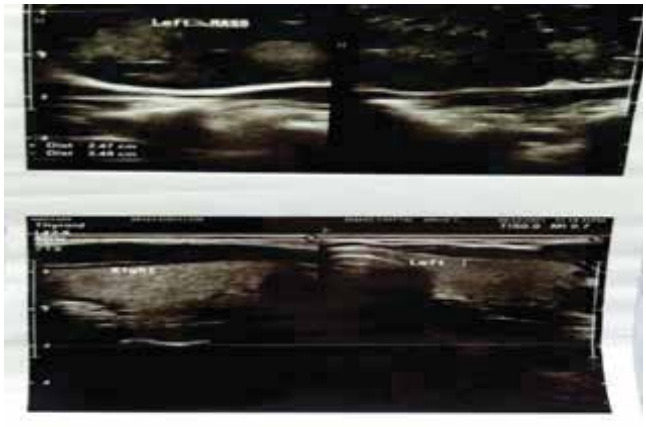

Figure-1: USG of the thyroid gland showing a solid nodule in the left lobe

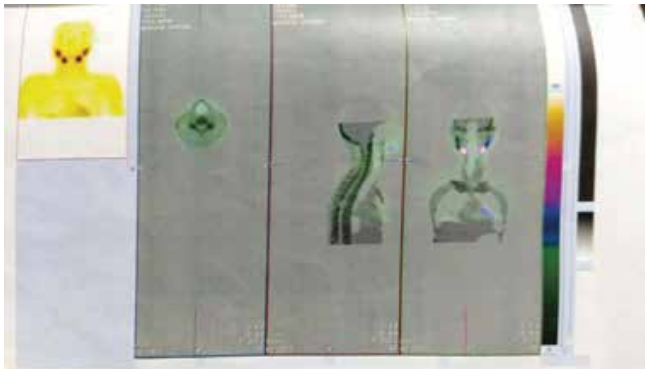

Figure-2: Parathyroid scan was negative for adenoma

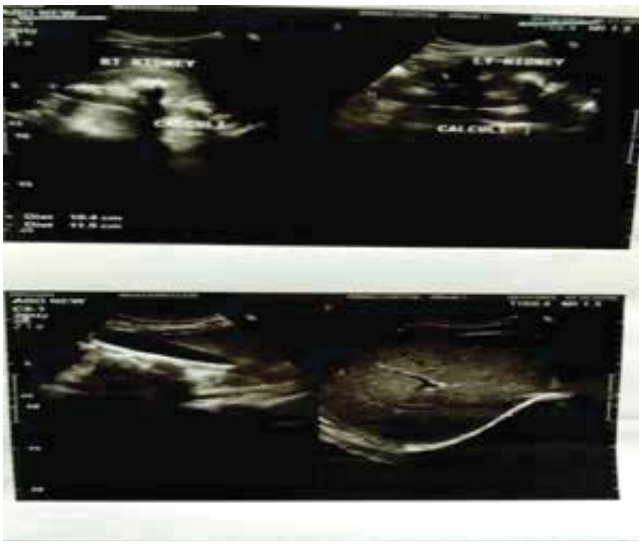

Figure-3: USG of Abdomen showing calcification in both renal pyramid

parathyroid mass was done under general anesthesia. Grossly a brownish mass was found that adheres to the thyroid lobe. The histopathologic appearance was that of a neoplasm, suggestive of parathyroid adenoma. Post-operative biochemical follow-up is represented in Table-I (1st follow-up). After 2-weeks, a medical board was arranged as the patient developed localized back pain. The board decided to do some investigation about 16 days after the operation (Table-I, 2nd follow-up). Whole-body bone scan suggested metastatic bone disease (Figure 4) and tissue from the metacarpal head (left) excision biopsy suggestive of osteitis fibrosa cystica. CT scan of neck and chest showed post-left hemithyroidectomy status and small enhancing soft tissue density structure about 10×8×7 mm in posteroinferior to lower pole of the right lobe of the thyroid - that suggests enlarged parathyroid gland.

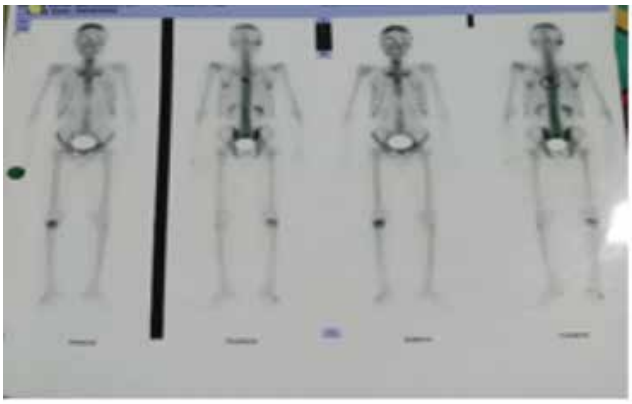

Figure-4: Bone scan of the patient showing metastatic bone disease

Immunohistochemical analysis demonstrated positivity to chromogranin A, GATA3 positive, and ki-67 (10% cells are positive) and suggested parathyroid carcinoma.

A decision was made for the completion thyroidectomy and excision of the remaining parathyroid gland. After the surgery, radiotherapy to the bony lesion and intravenous zoledronic acid with calcium supplementation was initiated. One month after surgery patient had a normal calcium profile with a normal parathyroid hormone (PTH) level (Table-I).

Discussion

Parathyroid neoplasm is rare, accounting for 0.4% to 5% of all cases of parathyroid hormone-induced hypercalcemia. It is the least common endocrine malignancy, with a prevalence of 0.005% of all cancers.[1-3] Parathyroid carcinoma typically presents between 45–59 years of age and occurs equally in men and women.[4] The pathogenesis of parathyroid cancer is unknown. It may occur sporadically or as a part of a genetic syndrome. Genetic syndromes that may have an association with parathyroid cancer cases include hyperparathyroidism jaw tumor syndrome (HPT-JT), MEN1, MEN2A, and isolated familial hyperparathyroidism.[4] Parathyroid carcinoma patients tend to show simultaneous manifestations of renal and skeletal involvement at the time of presentation.[4,5] Polyuria, renal colic, nephrocalcinosis, and nephrolithiasis are common renal complications. Bone pain, osteopenia, and pathologic fractures are manifestations of skeletal complications. Digestive complications include nausea, abdominal pain, peptic ulcers, and pancreatitis. Psychiatric symptoms include fatigue and depression.[4-6] Unless the patient presents with metastatic disease, a diagnosis of parathyroid carcinoma is usually only accomplished through a combination of intraoperative recognition of the disease and postoperative histological examination of the pathologic tissue. Therefore, the recommended approach for initial surgery with increasing diagnostic confidence is to obtain at least two localizing studies, most commonly a combination of a Technetium- 99m sestamibi scan and neck ultrasound. CT or MRI of the neck, mediastinum, chest, and abdomen may be beneficial for the determination of recurrence or metastatic spread.[4,5] However, FNAC can be useful in establishing the diagnosis of recurrent or metastatic disease by distinguishing scars from recurrent or metastatic parathyroid cancer tissue.[5,6] Laboratory

criteria to distinguish parathyroid adenoma from carcinoma are nonspecific. The average serum calcium level in patients with parathyroid carcinoma is higher (15 mg/dl) than that reported in patients with parathyroid adenomas (12 mg/dl). Only 10 percent of patients with carcinoma had serum calcium of less than 13 mg/dL and 75% had serum calcium of 14 mg/dL or higher.[5] Ki67 proliferation index is the most studied marker: it is higher in carcinomas (6–8%) than in adenomas (<4%), and a percentage greater than 5% generally suggests parathyroid carcinoma.[6] Complete surgical resection with microscopically negative margins is the recommended treatment and offers the best chance of cure. Parathyroid cancer has a high recurrence rate in up to 49–60% of cases after the initial operation. In cases of recurrence, surgical resection is still the primary mode of therapy.[4] Surgical complications include transient hypoparathyroidism, definitive hypoparathyroidism, hemorrhage, transient recurrent laryngeal nerve lesion, hungry bone syndrome, persistent hyperparathyroidism, and recurrence. Metabolic complications that can occur after resection include hypocalcemia and hypophosphatemia.[5,6] The aim of follow-up is early detection of potentially curable loco-regional recurrence and/or secondary tumors. It should include clinical examination with calcium levels and PTH monitored every 3 months for the first 3 years, 6 months till 5th year and yearly lifelong after that.[5]

Conclusions

Parathyroid carcinoma should be suspected despite its rarity when there is severe hypercalcemia, large cervical mass and concomitant renal and skeletal disease. Complete surgical resection at the earliest possible time is the optimal treatment to improve outcomes and offer the best chance of cure. Follow-up should be mandatory after surgery to improve quality of life.

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Conflict of Interest

The authors have no conflicts of interest to disclose.

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Data Availability

Any inquiries regarding supporting data availability of this study should be directed to the corresponding author and are available from the corresponding author on reasonable request.

Ethics Approval and Consent to Participate

Written informed consent was taken from the patient.

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