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Parathyroid Carcinoma presented with Renal Colic

Renal Kolikle Bulgu veren Paratiroid Karsinomu: Olgu Sunumu

Genel Cerrahi

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Özet

Paratiroid kanseri 1/1.000.000 'den daha az sıklıkta görülen nadir bir endokrin malignitesidir. Klinik bulgular, artmış parathormon ve kan kalsiyum düzeylerinin hedef organlardaki etkisine bağlı ortaya çıkmaktadır. Paratiroid kanserinin tedavisi en blok rezeksiyondur. Böbrek taşı şikayeti ile başvuran hastada yapılan tetkiklerde şiddetli hiperkalsemi (15.0 mg/dl) ve artmış parathormon seviyeleri (1520 pg/ml) saptandı. Hasta, yapılan boyun ultrasonografisi ve paratiroid sintigrafisi sonucunda paratiroid adenom ön tanısı alarak opere edildi. İntraoperatif frozen kesit tetkiki sonucunda paratiroid karsinomu tanısı alan hastaya ipsilateral tiroidektomi + paratiroidektomi + selektif boyun diseksiyonu yapıldı. Hasta 6 aylık rutin kalsiyum ve parathormon düzeyi kontrollerinde hastalıksız olarak takip altındadır. Paratiroid karsinomları çok nadir görülen endokrin malignitelerden olup, 15 mg/dl üzerindeki kan kalsiyum düzeyleri ve şiddetli hiperparatiroidi durumlarında akılda tutulması gereken, tedavisi cerrahi olan ve postoperatif dönemde nüks açısından rutin kontrolleri mutlaka yapılması gereken bir endokrin hastalıktır.

Anahtar kelimeler: *Paratriod karsinom*, *paratroid hormon*, *kalsiyum*

Abstract

Parathyroid carcinoma is a very rare endocrine malignancy with less than 1/1.000.000 incidence rate. Clinical findings are due to the effects of increased parathormone and blood calcium levels on target organs. The treatment of parathyroid carcinoma is en bloc resection. Severe hypercalcemia (15.0 mg/dl) and elevated parathormone levels (1520 pg/ml) weredetected in our patient who presented with kidney stone complaints. The patient was preliminarily diagnosed withparathyroid adenoma by neck ultrasonography and parathyroid scintigraphy. Ipsilateral thyroidectomy + parathyroidectomy + selective neck dissection were performed in the patient who was diagnosed as parathyroidcarcinoma by intraoperative frozensection analysis.Thepatient is under follow-up without any disease at the 6-month routine calcium and parathormone level controls. As a result, parathyroidcarcinoma is averyrareendocrinemalignancyand an endocrine disorder that should be kept in mind in cases with blood calcium levels above 15 mg / dl and severe hyperparathyroidism, which should be routinely controlled for postoperative recurrence.

Keywords: *Parathyroid carcinoma*, *parathormone*, *calcium*

Introduction

Only less than 1% of primary hyperparathyroidismcasesareduetoparathyroidcarcinoma (PTC)[1,2]. PTC is a veryrareconditionwith less than 1/1.000.000 incidence rate [3]. Primary hyperparathyroidism disease can be diagnosed very easily using a routine blood calcium test [4].

Theetiology of PTC is not veryclear. A rare autosomal dominant disorder(hyperparathyroidism – jawtumorsyndrome), familial isolated hyperparathyroidism, MEN-1 and MEN-2A [5] long standing secondary hyperparathyroidism and less clearly history of head and neck are possible predisposing factors.

The clinical features of PTC primarily are due to the effect of excessive secretion of parathyroid hormone by functioning tumor rather than to infiltration of vital organs by tumoral mass. The symptoms of PTC are similar to

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that of adenomas, but more severely presented, and most of the clinical manifestation are because of severe hypercalcemia.

Severe nephrolithiazis, nephrocalcinosis and impaired renal function (up to 80%), and severe bone involvement (up to 90%) are the main symptoms. Other symptoms of primary hyperparathyroidism are fatigue, loss of focus, malaise, bone pain, polydipsia, polyurea and depression.

Recurrent severe pancreatitis and peptic ulcer disease are common. A neck mass can be palpated in %22 of patients [6]. Hoarseness and palpable enlarged lymph nodes can also provide clue to the presence of a carcinoma.

There is no pathogonomic laboratory data, but in the presence of markedly elevated parathyroid hormone levels (>5 times the upper limits of normal) and serum calcium levels (usually more than 14-15 gr/dl) PTC should be suspected [7]. Severe hypercalcemia may not be observed in all patients.

The studies used for localization of parathyroid glands are ultrasonography, computed tomography, magnetic resonance imaging, sestamibi scan. Every technique has its own limitation. Once PTC is diagnosed, staging studies are not necessarily needed [1].

Case Report

A 33-year-old male patient was admitted to urology clinic of our hospital with complaints of hoarseness and previously diagnosed nephrocalcinosis. Routine laboratory tests revealed severe hypercalcemia (15.0 mg/dl) and elevated parathyroid hormone levels (1520 pg/ml). Thepatient was referred to our surgery clinic for further investigation. Ultrasonogaphy examination results revealed an adenoma like parathyroidmass (30x20 mm) at lower pole of the left thyroid lobe. The sestamibi scan revealed an intense activity at the area of lowerpole of the left thyroid lobe. Intraoperatively, a partially intrathyroidal parathyroid tumor was detected in the previously mentioned area and this mass was resected with ipsilateral thyroid lobe. The specimen was sent to the pathology clinic for frozen section examination. Upon the diagnosis of parathyroid carcinoma after frozen section examination a selective neck dissection was performed and lymphnodes from zones 2, 3, 4, and 6 were removed. Histopathological examination of total of 20 lymph nodes revealed no metastases. The patient is still free of disease after three years follow-up period.

Management

The only curative treatment of PTC is surgical resection. Since, incomplete excisions are associated with very high recurrence rates, en bloc resection at the time of initial operation provides thebest chanceforcure and long term survival. En bloc resection of PTC includes parathyroidectomy with ipsilateralthyroidlobectomy and removal of tracheoesophageal paratracheal and upper mediastinal lymphnodes.

Chemotherapy currently has a limited role in the treatment of PTC patients. Role of radiotherapy is promising but definitive conclusions are limited. Radiotherapy is also useful to treat unresectable recurrent neck disease or to palliate bone metastases.

There are 3 basic treatment aims in the management of hyperparathyroidism; to correct dehydration, enhance renal excretion of calcium, to inhibit accelerated bone reception. After treatment patient follow up must be done at 4-6 months intervals. Elevation of parathyroid hormon elevels during followup period is highly suggestive of recurrent disease. Factor sassociated with higher mortality rates include unadequate parathyroidectomy, positive lymph node

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Status anddistant metastases at presentation, and nonfunctional tumors.

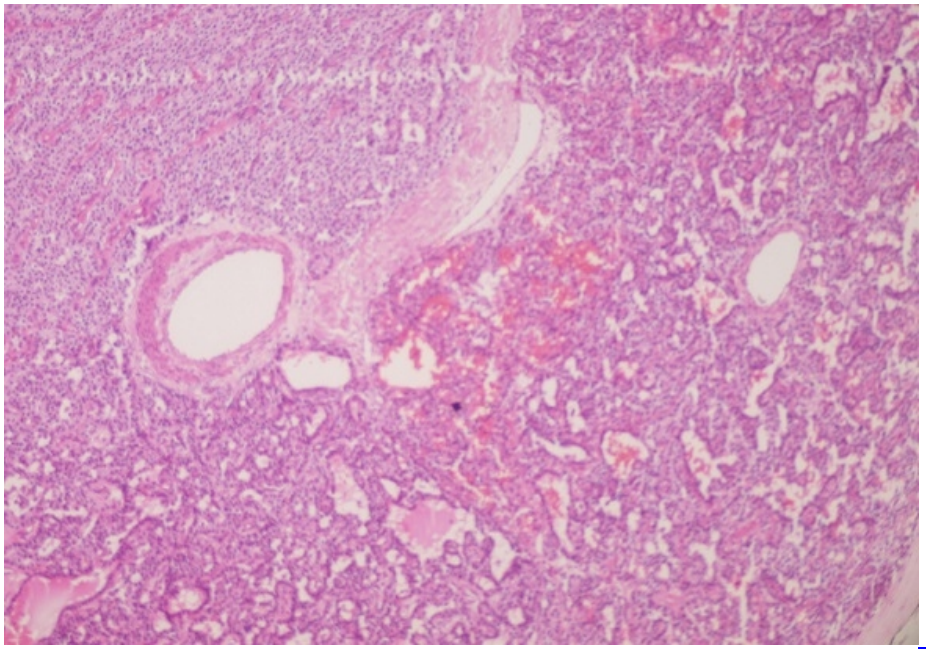

Figure 1
Hemotoxylin-eosin staining with 40X magnification.

Case Discussion

Parathyroidcarcinoma is a rare entity and often represented with symptoms like hypercalcemia, fatigue and palpabl neckmass in some cases although rarely the tumor may be non functional. Diagnosis can be challenging and novel biomarker slikegalectin-3 and HBME-1 may be used in discrimination of benign from malignant cases. Although tumour size greatert han 3 cm is associated with lymph node involvement en blocresection remains at the main stage of treatment. Hence R1 resection is unacceptabledue to possible tumorseeding. Data from small series suggested external beam radiation and chemotherapy could be possible adjuvant therapies somay help reducet here currencerates[8]. Yet there is not strongevidencefor it. In our case we first met incidentally with a parathyroidcarcinoma so we does not have enough experienceso selective lateral neck dissection was performed due to possible lymph node involvement. There is conflicted evidence therelations hipbetween lymph node involvement and overall survival[9]. Further research should be directed at identifying more effective therapies for this diseases.

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