

Incidental Non-Functional Parathyroid Carcinoma Identified During Thyroidectomy

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Parathyroid carcinoma is a rare clinical entity with fewer than 700 cases reported. PC accounts for 0.5–1% of all cases of primary hyperthyroidism, and its incidence is 0.005% of all registered cancers in the United States [1]. The common presentation of PC is symptomatic severe hypercalcemia including fatigue, bone pain, anorexia, memory deficit and parasthesias. Non-functioning parathyroid carcinoma is a very rare subgroup with only 16 cases reported [2]. Its diagnosis is more difficult due to lack of symptoms. Since the only presenting symptom is an expanding neck mass, it typically presents at an advanced stage [3]. We present a case history of a patient with an incidental finding of NFPC discovered during thyroidectomy for a combination of toxic adenoma and papillary carcinoma.

Patient Description

A 44 year old woman presented with a 15 year history of goiter. In the 2 preceding months the goiter progressively enlarged accompanied by weight loss and palpitations. Thyroid function tests revealed hyperthyroidism. The symptoms were treated successfully with mercaptopurine. Fine needle aspiration and ultrasound were inconclusive. Thyroid scan demonstrated a hot nodule in the left thyroid lobe and the patient was admitted for elective resection of the left lobe.

Physical examination was unremarkable aside from enlarged left thyroid lobe with a nodule measuring 3 cm in diameter. There was no palpable lymphadenopathy. Initial laboratory studies showed hyperthyroidism and the calcium level was 2.48 mmol/L (normal = 2.0–2.5).

At surgery, the right thyroid lobe appeared normal without nodules. The left thyroid lobe was enlarged 3.0 x 7.0 x 8.0 cm and was resected with the isthmus. The operative and postoperative course was unremarkable.

On pathology three nodules were identified. Two were thyroid nodules with papillary carcinoma evident in the smaller one. The third nodule was 1.5 cm in diameter and was defined as parathyroid carcinoma with vascular invasion. The patient underwent complete thyroidectomy and compartment lymphadenectomy. Pathology revealed no additional evidence of tumor tissue. The slides were reviewed by an eminent endocrine pathologist in the United States. The patient is followed yearly by ultrasoundography. At 5 years follow-up there is no evidence of recurrence.

Comment

The diagnosis of primary hyperparathyroidism has increased in prevalence in the past 20 years due to routine blood chemistry screening. Parathyroid carcinoma accounts for 0.5–1% of all cases of hyperparathyroidism [1]. The diagnosis of functional PC is often made early because of excessively elevated calcium levels accompanied by severe symptoms of hypercalcemia. The case of NFPC is different in many aspects. It is very rare, with only 16 cases reported [2], and its being asymptomatic explains why it is diagnosed in advanced stages with palpable neck mass and distant metastasis [3]. In the 14 cases reported since 1920 [2], the average age was 49, with a male-to-female ratio of 1.3:1. It was usually located in the left neck, as in our patient [4]. The tumor size ranges from 2.5 to 14.0 cm and our case is unique in its relatively small size, which contributed to its incidental discovery.

Functional PC is treated with en bloc resection. Adjuvant radiotherapy is recommended for patients at risk for local recurrence [5]. Chemotherapy does not seem to have a clinically significant effect [4,5].

Because of the scarce number of cases of NFPC there are no evidence-based recommendations for its optimal treatment or for the treatment of local recurrence or distant metastasis. Follow-up of NFPC is difficult because of normal parathyroid hormone and calcium levels and depends largely on clinical palpation and radiological imaging [5].

Despite adequate resection, NFPC carries a grimmer prognosis than functional PC [4], but this may be attributed to the late diagnosis of the NFPC. At 5 years after surgery our patient has no evidence of recurrent tumor.

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PC = parathyroid carcinoma
NFPC = non-functioning parathyroid carcinoma