

Parathyroid carcinoma: case report

Summary:

The authors present a case of parathyroid carcinoma in a patient with lateral neck swelling. Following a literature review, the clinical and diagnostic profile, treatment and prognosis of this rare disease are discussed.

Keywords: Cancer, Hypercalcaemia, Hyperparathyroidism, Parathyroidectomy, Recurrence

Introduction

Although parathyroid neoplasms are common and cause primary hyperparathyroidism, parathyroid carcinoma is a rare entity. Diagnosis and management are challenging due to the lack of clinical and pathological features that may reliably distinguish malignant from benign disease. The first known case, described by De Quervain in 1909, was a non-functional tumor whose malignancy was only revealed by the lesion's macroscopic features. In 1929 Wilder described a case in a patient with primary hyperparathyroidism admitted to the Mayo Clinic. In 1938 A The authors present a case of parathyroid carcinoma in a patient with primary hyperparathyroidism. Following a literature review, the clinical and diagnostic profile, treatment and prognosis of this rare disease are discussed.

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This cancer is responsible for 0.4–5.2% of cases of primary hyperparathyroidism. The 45–55 year age group is the most affected, with a slight predominance of cases in women

A case that came to our attention - the first in over 100 total thyroidectomies and very few parathyroid operations - prompted this paper and literature review to establish the current indications for the diagnosis and treatment of this rare disease.

Case report

The patient was a 52-year-old man with no family history of cancer. He was diagnosed with schizophrenia in 2012 and was under treatment with neuroleptic drugs.

In March 2019, thyroid ultrasonography revealed a slightly enlarged thyroid gland (rt) with normal morphology and "thyroid shows normal size and homogenous parenchymal echotexture. Gland margin are well defined. there is well defined hypoechoic solid suspicious lesion measuring about 2.9x2.0x2.6 cm & volume 8.45ml. colour Doppler shows high intra nodular vascularity another also quite well defined hypoechoic nodule predominantly solid measuring about 1.4x1.1cm seen in the rt lobe of thyroid gland. probably representing a benign adenoma, colour doppler shows no vascularity.

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Fig. 1

Well differentiated parathyroid adenocarcinoma. Stromal infiltration. (H&E, original magnification 200x).

FNAC shows, smears are markedly cellular. These reveal many follicular epithelial cells arranged in cluster, papilla like fronds, sheets and singly. The cells are relatively uniform having round to oval nuclei. Cells with frank anaplastic features are not seen. Pre-operative blood calcium levels remained normal (9.5 mg/dL; n.v. 8.2–10.4),

Patient was undergone for total thyroidectomy as suspected follicular lesion of thyroid preserving the parathyroids and RLN with the excision of extrathyroid mass.

histopathology report shows, section of the thyroid tissue show a neoplasm at one side. It is composed of uniform round cells, arranged in sheets and trabecular pattern. Remainder of the thyroid tissue are unremarkable.

The histologic appearance is consistent with parathyroid neoplasm, compatible with parathyroid adenoma. They advised for immunohistochemistry

Immunohistochemistry reports as, compatible with parathyroid carcinoma

patient was sent to oncologist, they search for distal metastasis. But not found

after for months of surgery, his serum calcium was 9.7 mg/dL, PTH was 118.5 pg/ml

Discussion

The main signs and symptoms of parathyroid carcinoma are due to high calcium and PTH levels. They include pathological fractures, joint and bone pain, fatigue, polyuria and polydipsia, kidney stones, muscular asthenia, nausea, vomiting, loss of appetite and weight loss.

These signs and symptoms are typical of primary hyperparathyroidism and also arise in patients who do not have a parathyroid carcinoma. They should therefore not be considered cancer-specific.

Diagnosis is more problematic in non-functional lesions, and the prognosis is worse. A visible or palpable lump in the front of the neck or ultrasound or CT evidence may give rise to the suspicion of parathyroid carcinoma. Where the lesion can be palpated, it appears as a hard, solid mass of from a few millimeters to some centimeters in size, strongly adherent to the thyroid and infiltrating the adjacent structures.

High serum calcium (>14 mg/dL) and PTH (especially when twice the normal value) are considered as indicative of carcinoma.

Diagnosis

In most cases, the suspected diagnosis of parathyroid carcinoma was reached on the basis of clinical signs and the finding of hypercalcemia, hypophosphatemia and elevated alkaline phosphatase and osteocalcin, markers of increased osteoclast activity. As already noted, the diagnosis is almost certain when there are particularly high PTH and calcium levels, which are unlikely to be seen in cases of benign hyperparathyroidism.

Although instrumental diagnosis is non-specific, ultrasound can reveal some signs of malignancy, such as echostructure, irregular margins, any pathological adenopathies and any invasion of the adjacent structures. Computed tomography (CT), magnetic resonance imaging (MRI), bone scintigraphy with ^{99m}Tc-sestaMIBI and bone scintigraphy and fine-needle aspiration biopsy (FNAB) can help confirm the diagnosis. However, the definitive diagnosis is provided by the pathologist.

On macroscopic examination, the tumor is hard; whitish with a very thick fibrous capsule. It is strongly adherent to the surrounding tissues. Capsule invasion is considered a sign of malignancy. Under the microscope, capsule, blood vessel and lymph invasion, stromal calcifications, fibrous trabeculae, enlarged nuclei and strong mitotic activity are considered to be signs of malignancy.

Treatment

Surgery is the gold standard for the treatment of parathyroid carcinoma. *En bloc* dissection of the tumor with the thyroid lobe, the ipsilateral parathyroid and any other affected tissue is the most suitable treatment and leads to the best prognosis. The radicalism of the surgery is important and it is essential to avoid damaging the tumor capsule, as any residual or dispersed cells could lead to a fast recurrence.

Sometimes it is possible to remove local recurrences. Laterocervical and central lymphadenectomy is generally carried out only if necessary.

Some authors consider radiotherapy to have some effect on preventing recurrences when used as a complementary treatment, while chemotherapy is agreed by all to be ineffective.

The treatment of parathyroid carcinoma aims not only to cure the disease but to obtain its biochemical remission: normalization of blood calcium and PTH levels, arrest of bone calcium depletion and regression of vascular, renal and neurological disorders. Continued high postoperative calcium and PTH levels are a sign of the disease's persistence (metastasis or residual disease). Follow-up involves periodic monitoring of calcium and PTH levels, markers for the disease's recurrence.

The outcome of surgery depends on a series of factors which together determine the prognosis. The most important factor is early diagnosis. However, the extreme variability of the biological behavior of each individual tumor should be borne in mind. Some relapse soon after surgery, especially locally, while others can remain dormant for several years before giving rise to distal metastasis. The presence and size of any locoregional metastasis at the time of surgery do not seem to influence the prognosis. Long-term survival is reported as 40–86% at 5 years and around 49% at 10 years. In the event of recurrence, survival at 5 years is 0% (9–13).

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Conclusions

Our case of parathyroid carcinoma was atypical of most cases reported in the literature. The literature review revealed that this type of cancer is rare, and usually occurs in people in their 40s. Many cases are sporadic, although familial hyperparathyroidism is a rare, autosomal dominant condition that is associated with an increased risk of cancer

Approximately 1–2% of cases of hyperparathyroidism are caused by parathyroid carcinoma. It is most commonly found in patients with primary hyperparathyroidism, but is less common in secondary or tertiary forms. Functional carcinomas produce parathyroid hormone, which causes hypercalcemia and signs of bone or kidney disease

The real incidence of parathyroid carcinoma is difficult to establish, due to variable diagnostic criteria. Its histological features are not specific, as they have also been found in adenomas or even local recurrences of benign tumors). For this reason, the tumor's malignancy must be unequivocally confirmed only after prolonged follow-up, bearing in mind both histological (capsular and vascular invasion) and clinical features (invasive appearance, recurrences, distal metastasis)