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Metastatic Medullary Carcinoma with Normal Calcitonin Level: A Reported Case

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Case Report

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Abstract: Calcitonin is a sensitive tumor marker for medullary thyroid cancer and is useful in preoperative diagnosis and postoperative surveillance for recurrent disease. Rare cases of medullary carcinomas with a normal or undetectable calcitonin levels have been reported in the literature. We present the case of a 46-year-old woman with a metastatic thyroid carcinoma non-secreting calcitonin.
Keywords: calcitonin; medullary thyroid cancer; procalcitonin; thyroid nodule.

INTRODUCTION

Medullary thyroid cancer (MTC) is a malignant proliferation of the calcitonin-secreting, parafollicular C cells of the thyroid. It is a rare cancer, accounting for approximately 3– 4% of all thyroid cancers in the United States, and it is the second most aggressive, after anaplastic thyroid [1].

Calcitonin, a polypeptidic hormone secreted by the neoplastic cells, is considered a very sensitive and specific MTC tumor marker**,** allowing diagnosis, follow-up, and prognostication of MTC [2]. Few cases of non-secreting medullary carcinoma calcitonin have been reported in the literature, posing a diagnosis problems and postoperative follow-up as is the case with our patient.

CASE REPORT

A patient aged 46; operated: lobo-isthmectomy for multi heterodular goiter with benign cytology with histopathological examination of a medullary thyroid carcinoma of 3 cm (fig-1); a calcitonin assay was carried out negative (3 ng / l).

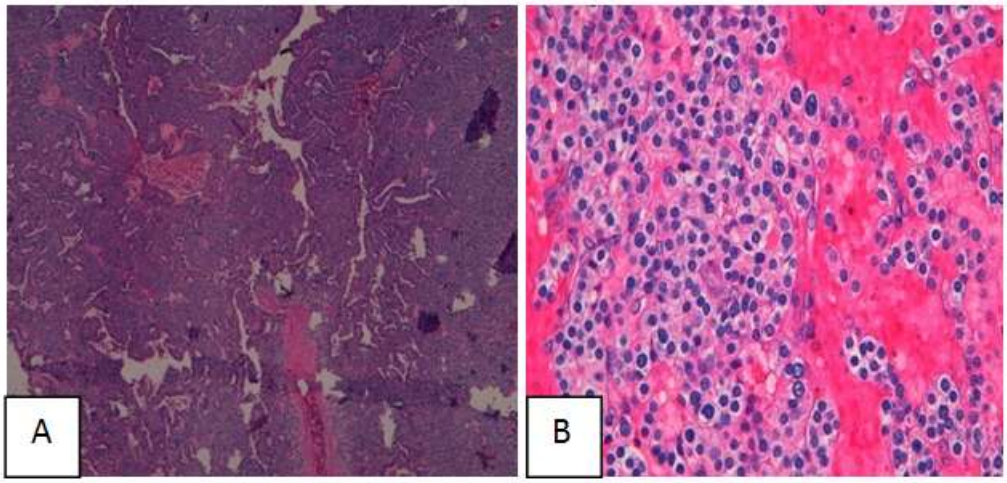

Fig-1: Surgical pathology of thyroid mass. A and B, Hematoxylin and eosin staining of the tumor at low power (A) and high power (B) showing solid architecture and cytological features consistent with MTC.

Totalising with lymph node dissection was performed returning without abnormality.

As part of the extension assessment, thoraco-abdomino-pelvic tomography was performed, demonstrating the presence of subpleural nodules and micronodules in the left hemi-pulmonary field, a left centromedullary iliac lesion and a peritoneal effusion of low abundance, corresponding to metastases.

An additional balance sheet: the carcinoembryonic antigen level was on the upper limit and pro-calcitonin was requested with the balance sheet of multiple endocrine neoplasia returning without particularity.

A complement of PET scan was requested but not realized by lack of means. The patient was followed by the encology

DISCUSSION

The medullary carcinoma of the thyroid has a biological specificity by the secretion of calcitonin allowing an early diagnosis, with a long-term follow-up [3, 4]. Spot cases of thyroid medullary carcinoma with a normal or undetectable calcitonin levels have been reported in the literature, their etiopathogenesis remains unclear. Some impute the degree of dedifferentiation of these tumors rendering their cells incapable of synthesizing calcitonin; while others link it to a defect of secretion that subsequently explains the low level of chromogranin and carcinoembryonic antigen in these cases [5-9]. Surgical approach thyroid medullary carcinoma with a normal or undetectable calcitonin levels does not differ from those secreting CT: surgical totalization with ganglion dissection, chemotherapy and metabolic radiotherapy [10, 11].

Postoperative follow-up of these rare cases should include periodical imaging: cervical ultrasound, magnetic resonance imaging or thoraco-abdomino-pelvic tomography associated with a bone scintigraphy or tomography, and measurement of all potential markers (carcinoembryonic antigen, calcitonin or procalcitonin). Patients with poorly differentiated MTC are at higher risk of disease-related death, and require more aggressive follow-up strategy [12-14].

CONCLUSION

Less than 1% of MTC are calcitonin-negative. It is even more unusual to find a cancer in which calcitonin is undetectable in the serum, especially in the presence of strong immunohistochemical staining for calcitonin. Possible causes for a lack of detectable calcitonin in the serum include ineffective post-translational processing or a defect in the secretion of calcitonin. Our case illustrates the challenges in managing a patient with calcitonin-negative MTC.

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