



Papillary thyroid carcinoma in a patient with MEN1 syndrome



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MEN1 is a rare syndrome characterized by hyperplasia or neoplasm of the parathyroid glands, pituitary, pancreas or duodenum and can associate less frequently pheochromocytoma, thymic or bronchial carcinoids, multiple lipomas, cutaneous angiofibromas and thyroid adenomas. Patients with untreated MEN1 have a decreased life expectancy, with a 50% probability of death by age 50 years

Case report: 47 year old woman with MEN1 presenting parathyroid recurrent adenomas, a pituitary prolactin-growth hormone cosecreting macroadenoma associated with an incidentally papillary thyroid microcarcinoma.

2002

Coraliform lithiasis surgically treated
Hypercalcemia
Increased PTH
Parathyroidectomy –
parathyroid adenoma

2004

Amenorheea
Hyperprolactinemia
Pituitary microadenoma
Starting dopamine agonist treatment

2014

No follow ups for 10 years
Another kidney stone was discovered
Hypercalcemia and increased PTH, normal TSH and free thyroxine levels.
Total thyroidectomy and parathyroidectomy. Besides the **two parathyroidian adenomas**, the histopathological examination revealed a **papillary thyroid microcarcinoma**.

One year after, she was admitted in our service. Checking her medical records we realize that we have a MEN1 patient, so we had to continue the investigation.

Paraclinic evaluation : Hematological and biochemical test with normal values.

Hormonal tests: Levothyroxine suppressive therapy for papillary thyroid carcinoma, FSH and estradiol adequate values for menopause, a slightly increased PTH level with normal calcemia most probably secondary to the vitamin D insufficiency, abnormal prolactin level and not expected increased IGF1 levels with inadequate suppression of GH in OGTT.

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|------------------------------------|-------------------------------|
| Prolactin | 36.97ng/ml (2.74-19.64 ng/ml) |
| IGF1 | 260ng/ml (156-217ng/ml) |
| GH in OGTT suppression test | 1.8mcg/l |
| Calcium | 9.7mg/dl (8.5-10.2mg/dl) |
| PTH | 70.3pg/ml (15-65pg/ml) |
| Vitamin D | 18.64ng/ml (30-100ng/ml) |

Pituitary CT scan revealed a moderately increased size of pituitary gland with right sided hypoattenuating nodule, measuring 1.01/1.03cm, with low enhancement after the contrast administration; no displacement of pituitary stalk. Additionally, the patient had no signs or symptoms suggesting a pancreatic-duodenal involvement (no hypoglycaemia and insulin and gastrin in normal range).

The next steps are the **genetic analysis** for MEN1 gene in our Endocrine Genetics Department and the treatment for the pituitary adenoma.

Conclusion: This case underline the different phenotypic presentation of MEN1. Our patient had the classical presentation hypercalcemia but she doesn't associate duodenoenteropancreatic NETs, the second most common endocrine manifestation in MEN1 syndrome. Additionally, the pituitary adenoma cosecrete prolactin and growth hormone. **As we found in literature, the papillary thyroid carcinoma is probably incidental. Mild symptomatology of this syndrome, low adherence to medical follow-ups and not having a clear medical report may be considered particularities of this case.**

References:

1. A Case of Multiple Endocrine Neoplasia Type 1 Combined with Papillary Thyroid Carcinoma Hai-Jin Kim, Jong-Suk Park, Chul-Sik Kim, Eun-Seok Kang, Bong-Soo Cha, Sung-Kil Lim, Kyung-Rae Kim, Hyun-Chul Lee, and Chul-Woo Ahn
2. Multiple endocrine neoplasia type 1: atypical presentation, clinical course, and genetic analysis of multiple tumors. Vortmeyer AO1, Lubensky IA, Skarulis M, Li G, Moon YW, Park WS, Weil R, Barlow C, Spiegel AM, Marx SJ, Zhuang Z.
3. Diagnosis and treatment of multiple endocrine neoplasia type 1 (MEN1). Gaztambide S1, Vazquez F, Castaño L.
4. Multiple endocrine neoplasia type 1. Marini F1, Falchetti A, Del Monte F, Carbonell Sala S, Gozzini A, Luzi E, Brandi ML.

