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
Amenorrhea
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 parathyroid 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.

Paraclinical 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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<thead>
<tr>
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<th>Value</th>
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<tbody>
<tr>
<td>Prolactin</td>
<td>36.97 ng/ml (2.74-19.64 ng/ml)</td>
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<tr>
<td>IGF1</td>
<td>260 ng/ml (156-217 ng/ml)</td>
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<tr>
<td>GH in OGTT suppression</td>
<td>1.8 mcg/l</td>
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<tr>
<td>Calcium</td>
<td>9.7 mg/dl (8.5-10.2 mg/dl)</td>
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<tr>
<td>PTH</td>
<td>70.3 pg/ml (15-65 pg/ml)</td>
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<tr>
<td>Vitamin D</td>
<td>18.64 mg/ml (30-100 mg/ml)</td>
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Pituitary CT scan revealed a moderately increased size of pituitary gland with right sided hypopattenuating nodule, measuring 1.01/1.03 cm, 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: