A case of multiple endocrine neoplasia syndrome type-2A (MEN-2A) with primary hyperparathyroidism and medullary carcinoma of thyroid initially treated by surgery is reported. The presence of pulmonary nodules six years after the initial treatment was thought to be related to metastasis of medullary carcinoma, and the increase in serum calcium concentrations was assumed to be caused by persistence of parathyroid adenomatous or hyperplastic tissue. The patient underwent surgery again and the pulmonary nodules were confirmed to be metastases of a parathyroid carcinoma, a very rare entity in MEN-2A syndrome.

Multiple endocrine neoplasia syndrome type-2A (MEN-2A) is an autosomal dominant disorder that comprises medullary thyroid carcinoma and variable prevalences of hyperparathyroidism and phaeochromocytoma. Hyperparathyroidism is usually caused by parathyroid hyperplasia and, rarely, parathyroid adenoma. Sporadic parathyroid carcinoma often presents as local invasion or recurrence, and a severe hypercalcaemia may be caused by distant metastases, particularly in the lung.1

CASE REPORT

A 49 year old man was admitted because of acute myocardial infarction. Serum total calcium concentration was 3.6 mmol/l, phosphorus 0.58 mmol/l, creatinine 168 µmol/l, and intact parathyroid hormone (PTH) 370 ng/l (normal range 10–55). Primary hyperparathyroidism was diagnosed. Cervical ultrasonography showed an image compatible with a parathyroid adenoma and a surgical neck exploration was performed. The left upper parathyroid gland was removed, and the pathological study revealed a 12.5 g parathyroid adenoma. Intraoperative biopsy of the inferior left gland showed the absence of fatty tissue; the upper right gland had a normal appearance and the lower right gland was not identified. After surgery, the calcium concentration was 2.67 mmol/l and PTH 118 ng/l.

Twelve months after surgery the patient underwent repeat cervical surgery for a thyroid nodule casually found in an ultrasonographic study. No pathological parathyroid tissue was found and a right hemithyroidectomy was performed. The pathological study showed a medullary carcinoma and the thyroidectomy was completed. Therefore, the patient was diagnosed with MEN-2A. Calcitonin was 75.8 ng/l (normal range 23–71) after surgery and 57 ng/l 17 months later. Metanephrines and vanillylmandelic acid concentrations were normal. Genetic study did not reveal any known mutation in the MEN-2A gene. Figure 1 shows calcaemia and PTH over time.

The patient didn’t have a known familial history of hypercalcaemia or any thyroid or adrenal neoplasia, but after this

**Abbreviations:** MEN-2A, multiple endocrine neoplasia syndrome type-2A; PTH, parathyroid hormone

![Figure 1](https://www.postgradmedj.com)

**Figure 1**  Concentration of serum total calcium during follow up. Dots represent PTH concentration. Normal range for calcium is represented between dotted lines. Arrows represent surgical procedures. (A) Upper left parathyroid gland removal. (B) Right hemithyroidectomy. (C) Total thyroidectomy. (D) Pulmonary segmentectomies.
bryonic antigen is well known in medullary carcinoma, adenomatous or hyperplastic tissue. Synthesis of carcinoem-
corten concentrations was attributed to growth of parathyroid static medullary thyroid carcinoma. The rise in calcium level. Usually MEN-2A associated hyperparathy-
rroidism is mild and develops after the diagnosis of medullary thoracic-turn 

Ten years after the initial diagnosis, serum calcium was 3.77 mmol/l, phosphorus 0.84 mmol/l, alkaline phosphata 365 U/l (normal range 56–119), and PTH 1399 ng/l. The patient developed a severe osteitis fibrosa cystica. It was thought that this was caused by a growth of hidden parathyroid adenoma-
thoracic-turn 

Another interesting issue in this case was the initial serum calcium level. Usually MEN-2A associated hyperparathy-
rroidism is mild and develops after the diagnosis of medullary thyroid carcinoma. The discordance between high PTH and normal calcium level after surgery in our patient may suggest a low bioactivity of the PTH molecule.

If we had suspected the real diagnosis we would have operated on the patient when the pulmonary nodules were discov-
ered, to remove parathyroid tissue and avoid or delay the bone disease. We think physicians need an open mind and should be aware that sometimes a patient can be “special”.

Learning points

- MEN-2A is an autosomic dominant disorder which comprises medullary thyroid carcinoma and variable prevalences of hyperparathyroidism and phaeochromocyto-

- Hyperparathyroidism is usually caused by parathyroid hyperplasia and, rarely, parathyroid adenoma. The presence the parathyroid carcinoma in MEN-2A is very unusual.

- Parathyroid carcinoma often presents as a local invasion or recurrence, and a severe hypercalcaemia may be caused by distant metastases, particularly to the lung.

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MEN-2A syndrome and pulmonary metastasis

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