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Ectopic Parathyroid Adenoma in the Framework of an MEN1

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Abstract: Multiple endocrine neoplasia type 1 (MEN1 or Wermer syndrome) is a rare hereditary disease, and especially its association with ectopic parathyroid adenomas is even more. Defined As a tumor developing in at least two endocrine glands including the anterior pituitary gland, the parathyroid glands and the duodeno-pancreatic endocrine tissue. This autosomal dominant hereditary pathology is caused by the mutation of the NEM1 gene encoding the tumor suppressor menin and located on chromosome 11q13. There is, however, sporadic cases account for 8 to 14%. Primary hyperparathyroidism is a frequent pathology secondary to autonomous overproduction of PTH (parathyroid hormone); it is mostly due to parathyroid adenomas (solitary or multiple). Diagnosis is purely biochemical. Cervical ultrasound and MIBI parathyroid scintigraphy is most often performed in the first line with good diagnostic performance, surgery is the only curative treatment for primary hyperparathyroidism, the reason why a preoperative localization and at best per-operative detection in the case of an ectopic adenoma to ensure proper resection and avoid subsequent recurrences.

Keywords: MEN1, Primary Hyperparathyroidism, Parathyroid Scintigraphy, MIBI, Surgery, Per Operative Isotopic Detection, SPECT/CT, Ectopy

1. Introduction

Primary hyperparathyroidism (PPH) is one of the most common endocrinopathies with a prevalence of approximately three in 1,000 people in the general population. It is due in 85% of cases to a secreting adenoma [1]. The parathyroid adenoma can be located in an ectopic situation and then represents a particular challenge both diagnostically and therapeutically. Here we review here the diagnostic and therapeutic approach of the primary hyperparathyroidism, about a complex clinical case associating an MEN 1 and the presence of a mediastinal parathyroid adenoma [2, 3].

2. Clinical Case

This is a 47-year-old man known since 2015 for an MEN 1 associating a pituitary micro adenoma of 4.5 mm, non-secreting, a right adrenal nodule of 28 mm, suspicious on the scanner, non-secreting, a cephalic pancreatic nodule of 12 mm and a lung mesenchymal tumor at the level of the LSG related to a desmoid tumor. Symptomatic and recurrent primary hyperparathyroidism, already operated on three times without success. This hyperparathyroidism is complicated by recurrent bilateral renal lithiasis and severe chronic renal failure.

Among his personal history we note hypertension (high blood pressure) followed since 2016 under triple therapy: Aprovel 300 mg/d, Moxonidine 0.4 mg 2*/d, Amlodipine 5 mg/d. there was no notion of MEN or other autoimmune pathologies in the family.

The onset of the disorders dates back to 2011, i.e. at the age of 37, marked by the occurrence of low back pain with hematuria, motivating the patient to consult a urologist, an abdomino-pelvic ultrasound was done objectifying a right adrenal adenoma following which the patient was referred to endocrinology where, after biological and morphological investigations, the diagnosis of MEN 1 was made in 2015.

The problem therefore began in 2015 when hypercalcemia was highlighted, accompanied by hypercalciuria and a frank increase in PTH to 3 times the upper limit of normal (exact values not available). The diagnosis of primary hyperparathyroidism is then made. We are not aware of the results of the first localization assessment carried out at the time, but we know that the patient

underwent two surgeries a year apart. These two interventions do not lead to remission of hypercalcemia.

In 2021, given the persistence of symptomatic hyperparathyroidism, a new assessment returned in favor of hypercalcemia hyperparathyroidism.

Calcemia (Ca) = 2.90mmol/l, Albuminemia (Alb) = 47.4 g/l, VitD =39.6 ng/ml

Calciuria of 24h= 64.91/24h, phosphaturia= 336.3 mg/24h, PTH= 202 pg/ml)

Bone mineral densitometry shows cortical osteopenia and trabecular osteoporosis with partially altered bone texture.

Femoral cortical bone Z score -2

Trabecular bone L1-L4 score -2.7

To this end, a new location assessment is carried out. A cervical ultrasound performed returning without abnormalities or visualization of the adenoma. MIBI-Tc99m scintigraphy was performed according to a dual-phase protocol objectifying a focus of hyperfixation of the radio-tracer in the right basilobas corresponding to a parathyroid adenoma already described on a CT-scan of 21*5*12 mm of major axes in retro-oesophageal. The patient was operated a third time in visceral surgery where a PTH assay was done before excision of the adenoma returned to 300pg/ml and post excision returned to 219pg/ml. The anatomopathological study of the surgical specimen returned in favor of a morphological appearance compatible with a parathyroid adenoma, a dosage made 2 days postoperative of the PTH returned to 261 pg / ml and a calcemia to 3.02 announcing the failure of the third surgical intervention.

The patient was re-addressed to our level for a parathyroid scintigraphy (SP) with MIBI-Tc99m where a protocol made of a dynamic acquisition 30 minutes and a late static acquisition 1H (figure 1 and figure 2) shows a focus of hyperfixation of rounded shape of the upper mediastinal which decreases in intensity at the late stage, the scintigraphic aspect was in favor of an ectopic parathyroid adenoma with a "rapid wash out", an acquisition coupled with the scanner (TEMP/CT) specified its retrosternal location (figure3).

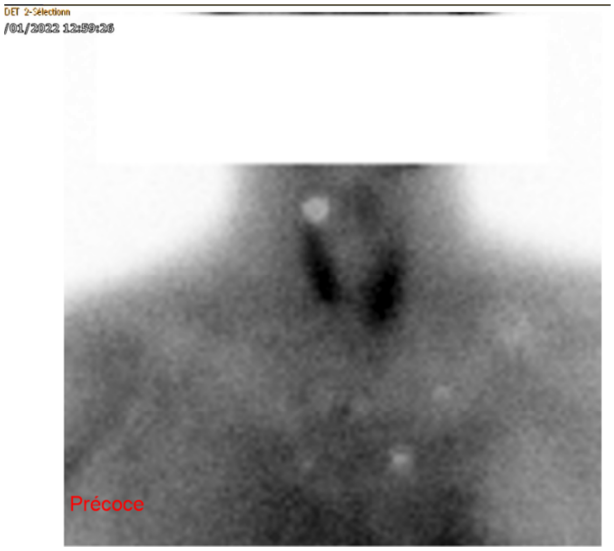

Figure 1. *Compressed dynamic acquisition.*

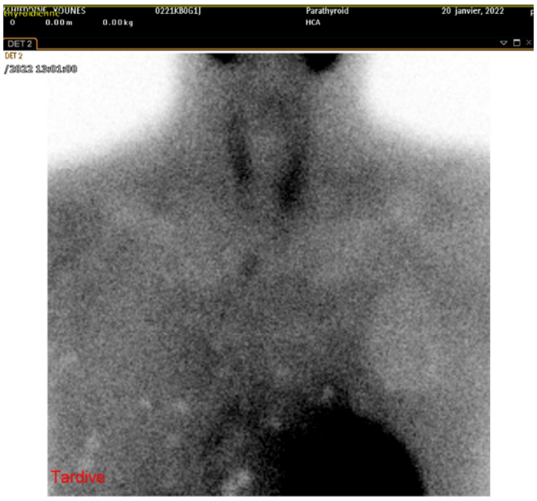

Figure 2. *Late acquisition.*

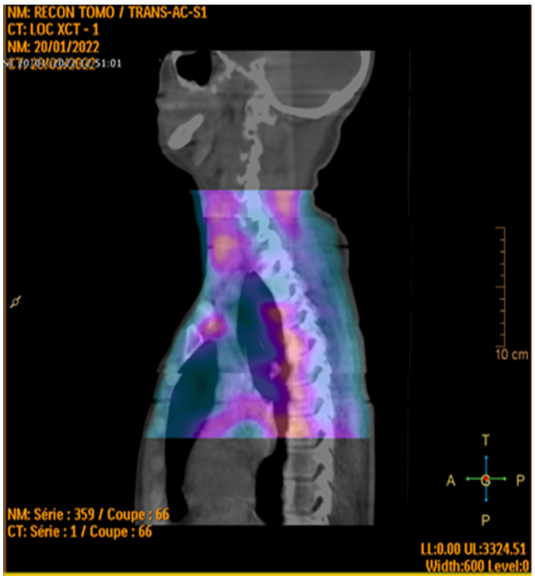

Figure 3. *SPECT/CT acquisition (retro sternal hyperfixation).*

The patient was presented for thoracic surgery for surgical excision of the adenoma under intra operative isotopic detection (DIPO).

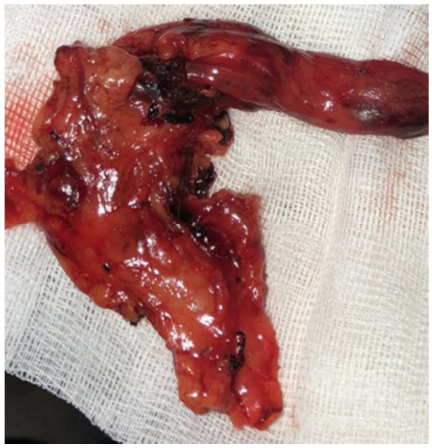

Figure 4. *Thymic parenchyma within which the ectopic parathyroid adenoma is located.*

Figure 5. *Intra operative isotopic detection device.*

The injection of 25mCi of the radiopharmaceutical MIBI-Tc99m was done in the operating room after the mid-thoracic incision, a PTH assay before the excision returned to 618pg/ml,

The detection probe (Figure 4) returned respectively: 700 counts per second (cps) after 30 minutes of injection; 572 cps at 1 hour post injection in vivo; 580 cps in ex vivo (figure 5).

A post-resection PTH assay returned to 257.3 pg/ml, therefore a decrease of more than 50%.

With good postoperative follow-up, a PTH assay made 2 days postoperative returned to 121.7pg / ml and on postoperative day 6 returned to 103.4 pg / ml, a serum calcium which was at 2.84 mmol / l 2d preoperatively returned to 2.26 mmol/l at day 2 post-operative and 2.48 at 2 months post-operative.

The anatomopathological study of the surgical specimen confirmed the presence of a parathyroid adenoma within an unremarkable involuted thymic parenchyma.

3. Discussion

We report the case of a 47-year-old patient followed since 2011 for symptomatic primary hyperparathyroidism. He benefited several times from adjustments in order to highlight the source of the inappropriate secretion of the parathormone, and from 3 surgical interventions all ended in failure the persistence of the symptoms and biological disorders, It is clear that he should only operate with a certain diagnosis and if possible with a highly probable preoperative localization [4]. Localization examinations cannot be considered as diagnostic arguments as they can expose to errors when this diagnosis is not established with certainty.

The correct localization of the pathological glands before the intervention currently calls upon cervical ultrasound and SP at MIBI [5].

These two examinations are very technical-dependent both in their performance and in their interpretation. With regard to cervical ultrasound, the best teams admit its lack of sensitivity in the detection of ectopic lesions, particularly mediastinal or retro-esophageal lesions, as well as in multi-faceted lesions. glandular [6].

SP at MIBI is also technically dependent. Admittedly, whatever the protocol, it detects ectopia, the subtraction method has the advantage of distinguishing thyroid nodules from parathyroid fixations and detecting multi-glandular involvement [7].

SPECT/CT improves the sensitivity of the examination in terms of localization, it is of great help in directing the surgical act, recent studies demonstrate its superiority compared to SPECT for the localization of parathyroid adenomas, both eutopic and ectopics [8, 9].

The contribution of DIPO is indisputable, which made it possible to guide the surgical procedure and to locate the ectopic adenoma in vivo [10].

4. Conclusion

Surgery remains the treatment of choice for primary hyperparathyroidism and the two recommended first-line localization examinations are cervical ultrasound and parathyroid scintigraphy with MIBI. However, in the presence of a parathyroid adenoma located in an ectopic position, these examinations are often faulty and the first exploratory cervical surgery generally results in failure and the persistence of hypercalcemia. Before any re-intervention, a precise localization of the adenoma is desired and a cervico-mediastinal SPECT/CT acquisition can be very useful, as well as the DIPO which will guide the surgical act.

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