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Parathyroid Carcinoma Clinical Presentation

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Summary: *Parathyroid carcinoma is involved in less than 1% of primary hyperparathyroidism. Parathyroid carcinoma patients usually present with striking hyperparathyroidism and hypercalcemia, with the resultant related symptoms being more severe than those associated with parathyroid adenomas. Parathyroid carcinomas also tend to be large and may be detectable by current imaging techniques. Its treatment is surgical, however when it is recognized in pre or intraoperative, carcinological surgery can be performe*
This study report the clinical and paraclinical cases of parathyroid cancers hospitalized in our department.
Keywords: *Parathyroid carcinoma, carcinological surgery, primary hyperparathyroidism.*

I. Introduction

Parathyroid carcinoma is involved in less than 1% of primary hyperparathyroidism.Parathyroid carcinoma patients usually present with striking hyperparathyroidism and hypercalcemia, with the resultant related symptoms being more severe than those associated with parathyroid adenomas. Parathyroid carcinomas also tend to be large and may be detectable by current imaging techniques. Its treatment is surgical, however when it is recognized in pre or intraoperative Its treatment is surgical, however when it is recognized in pre or intraoperative, carcinological surgery can be performe (1)(2).

The aim of this study is to analysis of the clinical and paraclinical cases of parathyroid cancers hospitalized in our department .

II. Obervation

Patient aged 29 years, hospitalized for management of a severe osteomalacia: bone pain, paresthesia, functional impotence of the inferior limbs with dandinante gait, complicated vertebral compaction at the origin of a cyphoscoliosis and pathological fracture of the femoral neck .

Radiography of the skeleton shows a diffuse demineralization and appearance of brown tumors in the bones of the hand . Cervical palpation found a very firm left lobar nodule. The cervical ultrasound demonstrated a left thyroid lobe increased volume of seat of a hypoechoic nodule of 25/29 mm whose cytopunction returned in favor of a vesicular carcinoma.

The diagnosis of hyperparathyroidism was established with very high PTH levels 1210 pg / ml (15-65) compared with hypercalcemia at 140mg / l (85-110). The myoview scintigraphy found a toto left lobar fixation.

The intraoperative exploration found a left intra-thyroid nodule. The patient underwent a total thyroidectomy with homolateral lymph node dissection and removal of upper left parathyroid

and lower right. The anatomopathological study found a parathyroid carcinoma infiltrating the thyroid capsule with ganglion metastasis. The parathyroids were hyperplastic.

The patient presented postoperative severe hypocalcemia treated with parenteral substitution. Thereafter serum calcium and PTH levels normalized.

The extension report and the reassessment made regularly during the following 7 years did not show any recurrence

III. Observation

Patient 42 years hospitalized for suspicion of hyperparathyroidism with a clinical picture of renal and bone evolving during a year in a context of emaciation and alteration of the general state There were recurrent bilateral renal colic complicated of bilateral nephrocalcinosis, diffuse bone pain, paraesthesia of the lower limbs with dandinizing gait, Vertebral collapse and cyphoscoliosis.

Cervical palpation found a lower right nodule. Cervical ultrasound showed a right basi-cervical nodule of 35 / 40mm. Hyperparathyroidism was confirmed with elevated PTH levels at 2279 pg / ml (15-65) facing major hypercalcemia at 179 mg / l (85-110) requiring several days of rehydration Peroperative exploration reveals a 6 cm nodule of whitish appearance with haemorrhagic changes infiltrating the thyroid and thymus

The patient received a lobo-isthmectomy right taking parathyroid nodule with removal of both left parathyroid and thymus. The anatomopathological study confirmed the parathyroid carcinoma with oxyphilic cells infiltrating the thymus with thyroid capsular effacement; The two parathyroids were hyperplastic.

The patient presented postoperative hypocalcemia treated by parenteral substitution. Subsequently the phosphocalcic balance and PTH normalized.

The extension assessment, the reassessment made during the next 3 years, was normal.

IV. Obervation

45 years old patient consulted for exploration of a left neck mass discovery farm in self-examination two months earlier. The cervical ultrasound finds a left basilobar nodule of 35/29 mm containing micro calcifications with central and peripheral mixed vasculature. Hyperparathyroidism was confirmed by elevated PTH levels at 249 pg / ml (15-65) for normal calcemia with vitamin D deficiency. The scintigraphy with the MIBI sesta finds a fixation corresponding to a left basicervical parathyroid Adenoma without other mediastinal localizations.

Intraoperative exploration found a 4 cm tumor developed at the expense of the body of the thyroid, adherent to préthyroïdien muscle and infiltrating cell. The patient underwent a block gesture involving the thyroid with central lymph node dissection. The anatomopathological study concluded a malignant tumor infiltrating the thyroid capsule with vascular emboli without being able to cut between medullary carcinoma and parathyroid carcinoma.

The diagnosis of certainty was made after the immunohistochemical study. The postoperative course was uneventful, PTH levels returned to normal. The staging is reviewed without anomalie.Le patient was substituted by vitamin D.

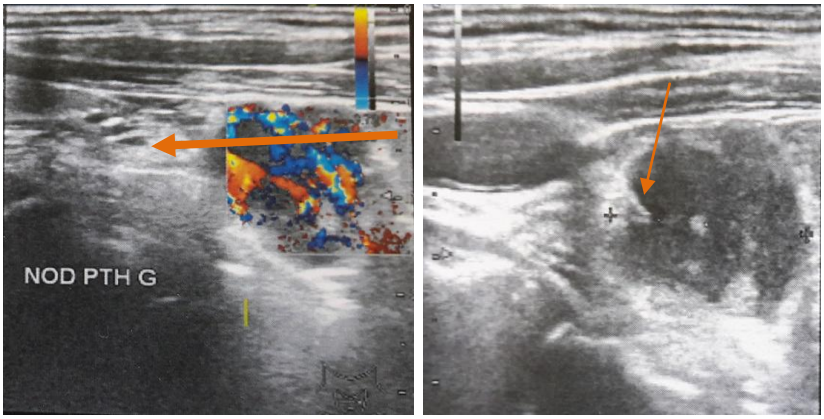

Fig 1 and 2: Vascularized hypoechogenic heterogeneous parathyroid nodule

V. Observation

Patient aged 47 was hospitalized for suspicion of hyperparathyroidism in front of a high calcemia found during a standard assessment. The cervical palpation has not found a nodule The hormonal balance confirmed hyperparathyroidism with very high PTH levels at 436.44 pg /ml (15-65), compared to asymptomatic hypercalcemia at 148mg / l without renal or bone damage.

Ultrasound showed an hypoechoic parathyroid nodule of 17mm with clear boundaries confirmed by MIBI scintigraphy with a fixation projecting behind the base of the left thyroid lobe. Intraoperative exploration found a nodule 20 /15mm with pretty clear limits, whitish appearance, important seat of above named inflammatory, adhering to the lower pole of the thyroid The anatomopathological study returned in favor of an adenocarcinoma of the parathyroid to Oxyphilic cells with blistering confirmed by a second proofreading.

The postoperative sequences were simple. Due to the break-in of the adenocarcinoma capsule and adherence to the thyroid lobe we completed with a total thyroidectomy in a second time. The 3-month, 6-month, and 1-year postoperative change resulted in normalized calcium phosphate and PTH levels, with no signs of recurrence or metastasis with the cervical ultrasound, toto-corporeal MIBI scintigraphy and the thoraco-abdomino-pelvic CT scan.

VI. Discussion

Primary parathyroid cancer is a rare clinical entity reported in the literature often in the form of limited clinical cases or series.

It is responsible for less than 5% of all primary hyperparathyroidism. Its histological diagnosis remains difficult in the absence of recurrence or distant metastasis; Its diagnosis is currently improved by the advent of immunohistochemistry (3)(4). His treatment is surgical. The quality of the first act opératoir influences the prognosis and survival. Even in the absence of consensus criteria, a strong pre or intraoperative suspicion should lead to a wide surgical gesture. The aim of this work is to try to clarify these elements of presumption (5).

Unlike parathyroid adenoma, parathyroid cancer affects men and women equally. The age is younger with an average towards the fourth decade. In our study the youngest of our patients was under 30 years old and the sex ratio was 1/1.(5)

The etiology of parathyroid cancer remains unknown. Usually, it is sporadic, family cases are reported. They are of autosomal dominant transmission and integrate or not in multiple endocrine neoplasias of type 1 or 2 NEM 1 -2 or other genetic diseases like the HJTS syndrome in connection with mutation of the HRPT2 gene .

The cervical irradiation was also incriminated. No personal or family history has been found in our patients. The existence of parathyroid hyperplasia in our patients may suggest that there may be an evolution from benign lesion to carcinoma(6)(7)

While parathyroid adenoma hyperparathyroidism is most commonly discovered by chance, the parathyroid carcinoma clinic is usually patent and severe (in more than 90% of cases), due to high levels of resistant PTH, Symptomatic hypercalcemia with severe forms: bone, renal or major hypercalcemia crisis with other signs of hypercalcemia.(8)

Asymptomatic forms are rare but may exist as evidenced by the case of our third patient with normal calcemia which may be explained by a previously negative calcium balance due to vitamin D deficiency and the fourth patient who was totally asymptomatic despite the high serum calcium level. He was probably diagnosed early before the installation of complications of hypercalcemia (9)(10).

Unlike parathyroid adenoma, the high PTH level reflects cell mass in the case of parathyroid cancer. This explains the palpation of the cervical mass in 30% to 70% of the cases according to the series. 3 of our patients had a palpable cervical mass element that should arouse suspicion in front of major signs of hypercalcemia (9).

The three patients had tumors greater than 2 cm neck ultrasonography (average 3cm), which is reported in the literature.

Another adenoma and carcinoma differentiating factor is the fact that the adenoma is not palpable whereas the tumor size is consistent (case of the fourth patient in whom the adenoma was not palpable despite a size of 2 cm). Ultrasound may also find suspicious but non-specific signs such as hypoechoic, mixed vascularization and micro calcifications. It can also underestimate the size of the tumor as the case of the second patient described at 4cm then what was actually at 6cm (11)(12).

When ultrasound does not allow the distinction between a thyroid nodule and a parathyroid adenoma especially when it is developed at the expense of the thyroid, MIBI scintigraphy makes it possible on the one hand to make the diagnosis and on the other hand research of the ectopic or metastatic foci even if it does not make it possible to differentiate a carcinoma and an adenoma (12)

The biological diagnosis of hyperparathyroidism is not a problem. PTH levels are severely higher compared to parathyroid adenoma. They can achieve in some descriptions 10 times the standards.

In parallel ,hypercalcemia is greater than 140mg / l or 30 to 40 mg more than the upper limit . Our two symptomatic patients had PTH levels 20 and 35 times the standard in the bone form and an average of serum calcium at 135 and 153 mg / l

In peroperative some elements may evoke the diagnosis of parathyroid carcinoma if it was not suspected preoperatively: the size of the tumor greater than that of the parathyroid adenoma beyond 2cm, the hard consistency, the color The presence of inflammatory reaction, the intimate adhesion to the thyroid or even the thyroid capsule intrusion and the infiltration of the neighboring organs.(1)(2)(13)

In our series, the macroscopic aspect of the tumor was whitish and adherent to the thyroid both in the two patients in whom the diagnosis of cancer had been suspected preoperatively as in the other two asymptomatic patients.

In the third patient the diagnosis was evoked in peroperative thanks to the experience of the surgeon due to the hemorrhagic character and the infiltration of the neighboring tissues.

The prognosis of our patients was good in the short and medium term, probably because of the extent of the surgical procedure.

At the end of this analysis, the clinical and paraclinical elements reproduced in Table I (in accordance with the data from the literature) which make it possible to strongly suspect the diagnosis of parathyroid carcinoma before surgery to indicate at the outset a broad surgical gesture rather than a minimal gesture that should be reserved for benign primary hyperparathyroidism

Table I: Clinical and paraclinical elements of presumption

	literature		Our series				
	Parathyroid adenoma	Parathyroid Carcinoma		Patient 1	Patient 2	Patient 3	Patient 4
Average age (years)	55	44-54	37	29	42	45	47
Sex ratio M/F	5/1	1/1	2/1	♀	♀	♂	♂
Clinical							
Palpation of the nodule	Rare <2%	Commun 38%	¾	Yes	Yes	Yes	Yes
Asymptomatic	40-80%	< 2%	50% ½	-	-	+	+
Bone shape	<5%	34-91%	25% ¼	+			
kidney shape	<18%	32-80%	0%				
Mixed form	Rare	Common	25% ¼		+		
Ultrasound Size nodule cm	< 2	>3	≥ 3 ¾	2,5/3	3,5/4	3,5/2,9	20/1,5
Calcemia mg/l	<120	> 140	156	140	179	normale	148
PTH pg/ml	↗	↗ ↗↗	↗↗ ↗	1210	2279	249	436
Macroscopy (Per operative)	Soft, oval, Red brown Without infiltration	Firm, gray whitish, lobulated, fibrosis, inflammation, adherence to the thyroid, Infiltration of neighborhood	Blanched, sclerosis, Intrusion of the capsule adenoma, Adherence to the thyroid, Neighbor infiltration in our 3 patients				

VII. Conclusion

If the biological diagnosis of hyperparathyroidism is easy, the histological diagnosis of parathyroid carcinoma remains difficult outside metastasis. The prognosis depends on the quality of surgery.

In the presence of primary hyperparathyroidism, the existence of certain clinical and para-clinical elements, makes it possible to suspect a parathyroid cancer in preoperative by the clinician and in peroperative by the surgeon

This should involve a broad carcinological surgical gesture rather than a minimal gesture even in the absence of consensus criteria Indeed, only an adequate curative treatment can improve the prognosis of the patient.

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