

EXTREMES IN HYPERPARATHYROIDISM: 'management of parathyroid carcinoma'

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Parathyroid carcinoma (PC) is a rare cause of primary hyperparathyroidism (PHPT), accounting for less than 1 %, with an equal gender distribution and an average age of diagnosis in the fifth decade of life. The diagnosis of PHPT is based on the laboratory finding of high levels of immunoreactive PTH in the presence of severe hypercalcemia. [1] Histopathological features of PC can overlap with those of adenoma, and is of great importance to consider the overall clinical and pathological picture. [2]

The only potentially curative treatment for PC is surgery. Early surgery is the most important factor for optimal outcome. [3] Despite the general belief that PC is radio-resistant, several series did report reducing local recurrence rate using external radiotherapy immediately after surgery. [4] No specific chemotherapy is established, due to the PC low incidence and different schemes applied.

PC has a variable prognosis, with a high rate of local recurrence and distant metastases. The 5-year overall survival is about 85.5 % and 10-year survival is 49.1 %. [5]

This case series is based on a multidisciplinary review of four patients with parathyroid cancer, describing their therapeutic management and follow-up !

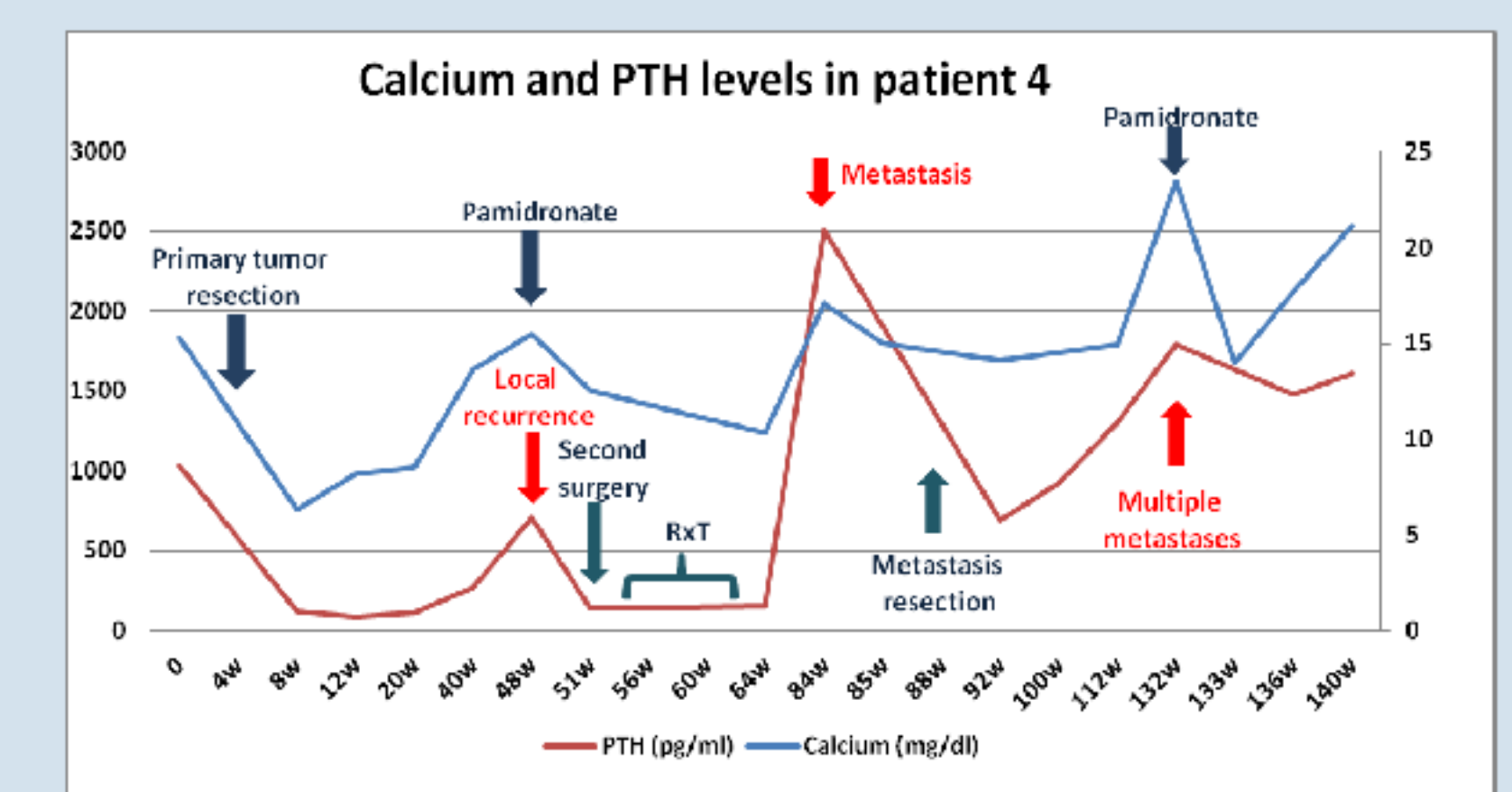
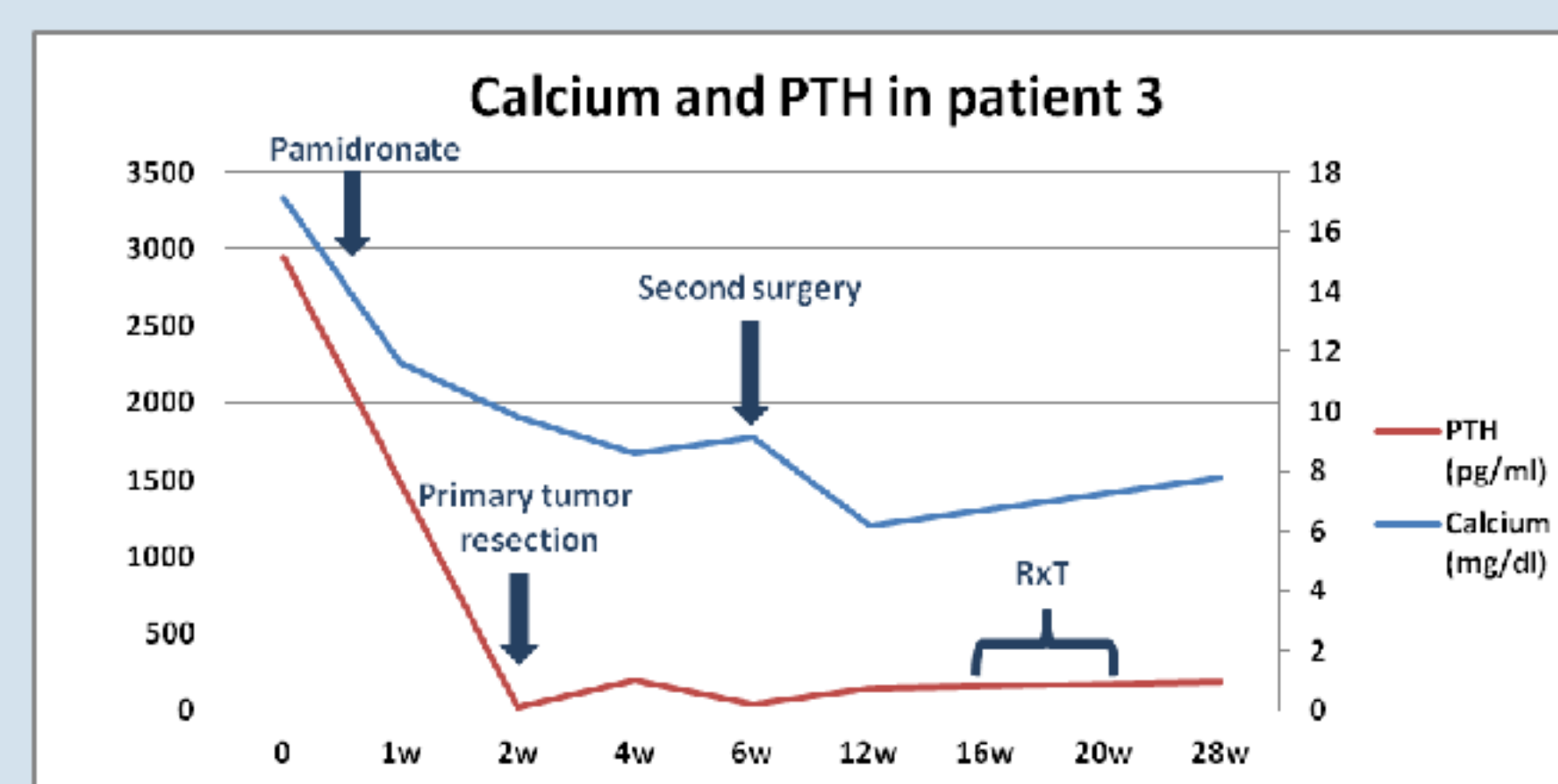
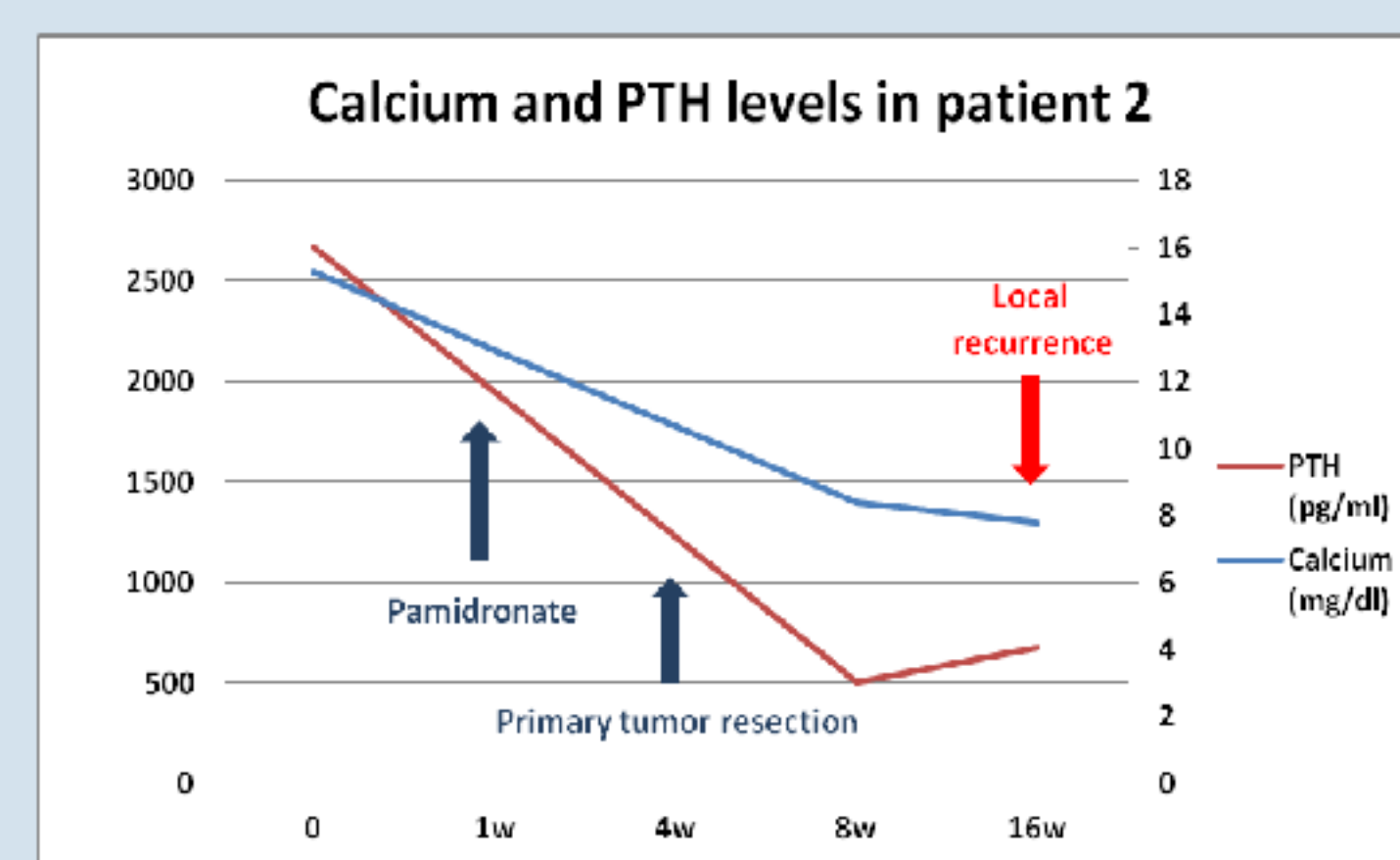
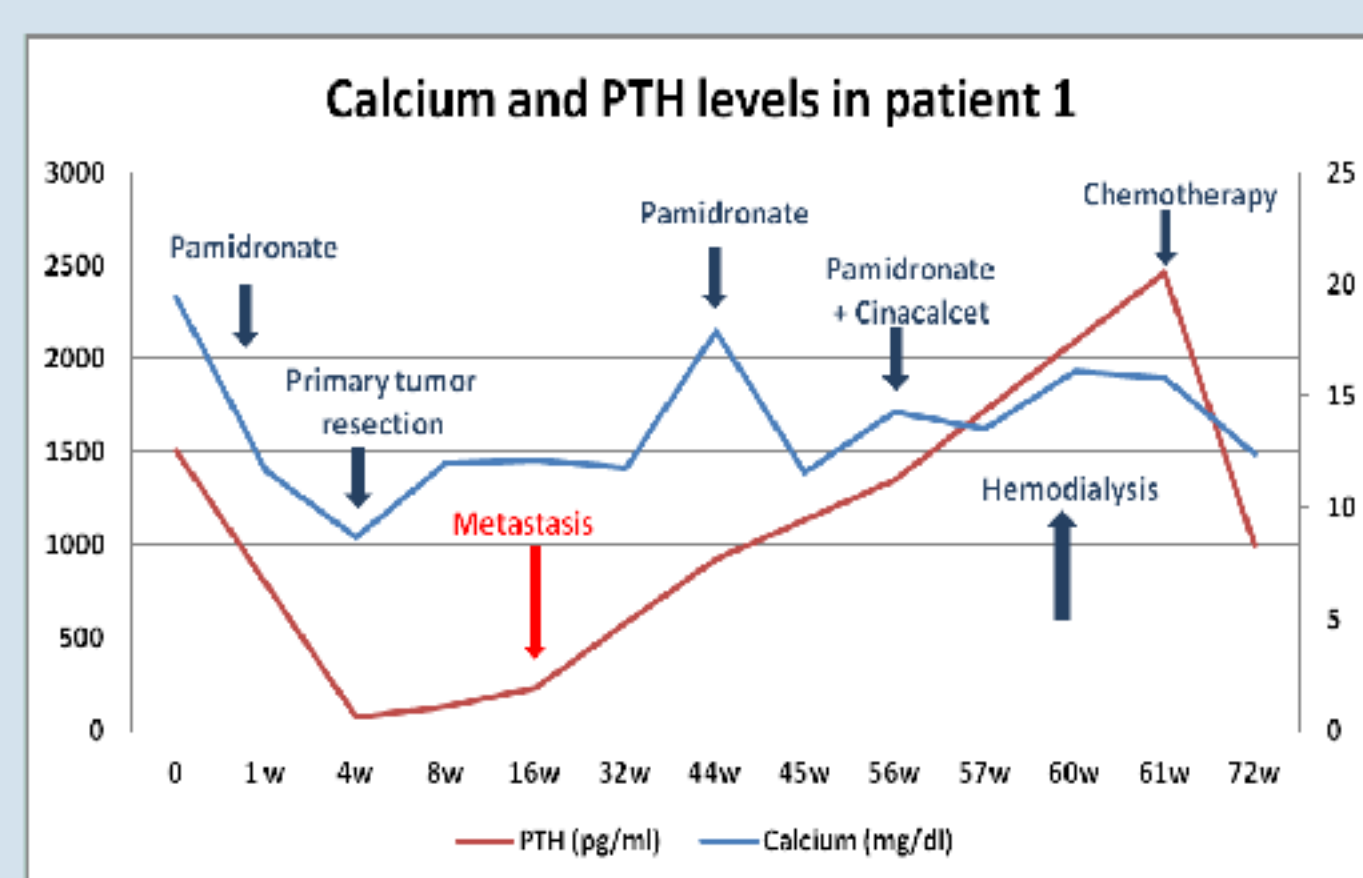
Between 2008 and 2014 four patients were diagnosed with PC in our clinical department, three men and a woman, with a mean age of 50 years \pm SD 13.22 (range 38-68). All had severe hypercalcemia (15.3-19.4 mg/dl) and elevated PTH levels ranging from 15 to 45 times above normal value. Tumor size ranged from 3.2 to 7 cm; two of them had thyroid gland invasion and one thymic invasion. Three patients underwent parathyroidectomy with hemithyroidectomy and one underwent parathyroidectomy with thymectomy and cervical dissection. Schulte stage at diagnosis was between II and IV, while all were classified as high risk.

Conformational radiotherapy of the tumor bed was used in 2 cases. Cinacalcet treatment was tried in one case and chemotherapy regimen in another, without significant improvement.

Recurrence occurs in approximately 50 % of cases, and is biologically detected by elevated serum calcium and PTH levels. In our case series two patients had local recurrence and the time from the initial surgery to recurrence ranged from 1 month to 1 year. Three patients were diagnosed with metastatic disease.

	Patient 1	Patient 2	Patient 3	Patient 4
Age (years)	68	38	53	43
Gender	Female	Male	Male	Male
Serum calcium (mg/dl)	19.4	15.3	17.1	15.3
PTH above normal level	23 x NL	41 x NL	45 x NL	15 x NL
BMD distal radius (g/cm ²)	0.400	0.282	0.505	0.367
Tumor site	Left inferior	Left inferior	Left inferior	Right inferior
Tumor size (cm)	3.2	7	4	3.7
Thyroid gland invasion	+	-	-	+
Lymph node invasion	+	+	+	+
Type of resection	Parathyroidectomy Hemithyroidectomy	Parathyroidectomy Thymectomy	Parathyroidectomy Hemithyroidectomy	Parathyroidectomy Hemithyroidectomy
Stage according Schulte differentiated classification at diagnosis	Stage III	Stage IV	Stage II	Stage II
Stage according Schulte risk classification	High risk	High risk	High risk	High risk
Local Recurrence	-	+	-	+
Distant Recurrence	Pulmonary	Mediastinal	-	Pulmonary, pleural, pancreatic, renal, suprarenal, bone
Radiotherapy	-	-	+	+
Chemotherapy	+	-	-	-
Period of follow-up	1 year and 6 months	3 months	8 months	4 years
Time between surgery and the first recurrence	1 month	2 months	-	1 year
Death due to PC				+

Table 1. Clinicul and pathological features of patients



In the graphics above we present the 4 cases of PC with different therapeutic management and the biochemical response based upon values of PTH and serum calcium.

CONCLUSIONS:

Parathyroid carcinomas are rare endocrine cancers, with high relapse rate and poor prognosis.

Multidisciplinary approach requires detailed imaging, skilled surgeons, endocrinologist and oncologist.

References

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- [2] Villar-del-Moral, Jesús, et al. "Prognostic factors and staging systems in parathyroid cancer: A multicenter cohort study." *Surgery* 156.5 (2014): 1132-1144.
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