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Occult Neuroendocrine Tumor Causing Severe Hyperparathyroidism

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INTRODUCTION

- We describe the diagnosis and successful management of a rare case of occult severe hyperparathyroidism due to a well-differentiated pulmonary neuroendocrine tumor.
- While hypercalcemia improved after tumor resection, the patient developed hungry bone syndrome causing hypocalcemia.

CASE PRESENTATION

- A 39-year-old female who presented with leg pain and no other symptoms was found to have a 9-cm lytic femur lesion and 3-cm right lower lung lesion.

Lab	Result	Normal Range
Calcium	17.5	8.6-10.0 mg/dL
Albumin	3.4	3.6-5.1 g/dL
Creatinine	2.17	0.6-1.2 mg/dL
Alkaline Phosphatase	766	31-125 U/L
PTH (non-diluted)	>2000	19-88 pg/mL
Vitamin D	11	30-100 ng/mL
PTHrP	17	14-27 pg/mL

- Hypercalcemia was treated with IV fluids, calcitonin, and IV pamidronate.
- Neck ultrasound did not show a parathyroid lesion.
- Intraoperative diluted PTH was 678 pg/mL and postoperative PTH 51 pg/mL following right lower lung lobectomy.

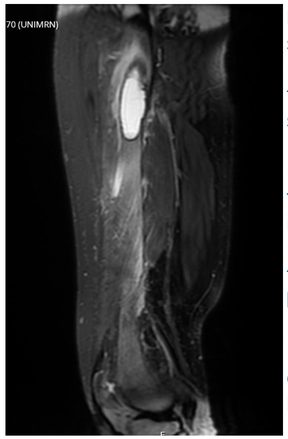

Figure 1. MRI Femur shows an expansile mass and innumerable tiny intracortical foci of signal abnormality. Pathology from open reduction and internal fixation showed increased bone resorption, peritrabecular fibrosis, and granulation-like tissue with hemosiderin, hemorrhage, and giant cells, compatible with bony changes of hyperparathyroidism.

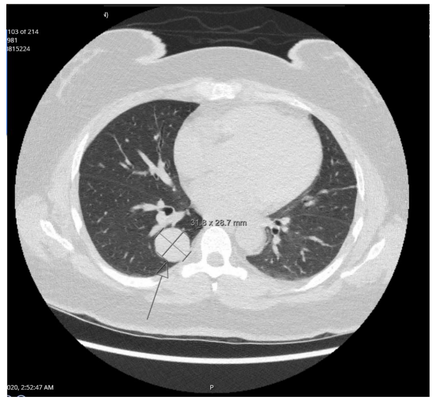

Figure 3. CT chest shows a 3-cm soft-tissue nodule in the right lower lung.

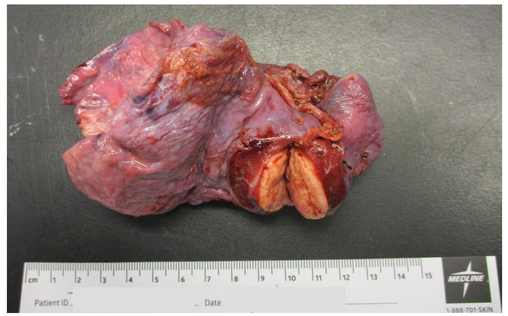

Figure 4. Gross pathology from right lower lung lobectomy. Pathology showed a benign 4 cm well-differentiated intermediate-grade neuroendocrine tumor with focal necrosis, 3-4 mitoses/10HPF, and 15% proliferative index with Ki-67 (Figure 5).

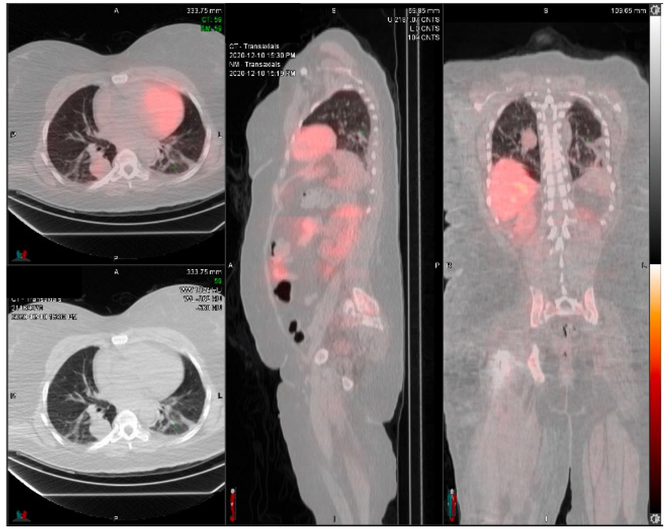

Figure 2. Whole body sestamibi T-99 scan showed no signs of a parathyroid lesion, but showed moderate uptake in a round mass in the right lower lung and intense uptake in the fundus of the stomach.

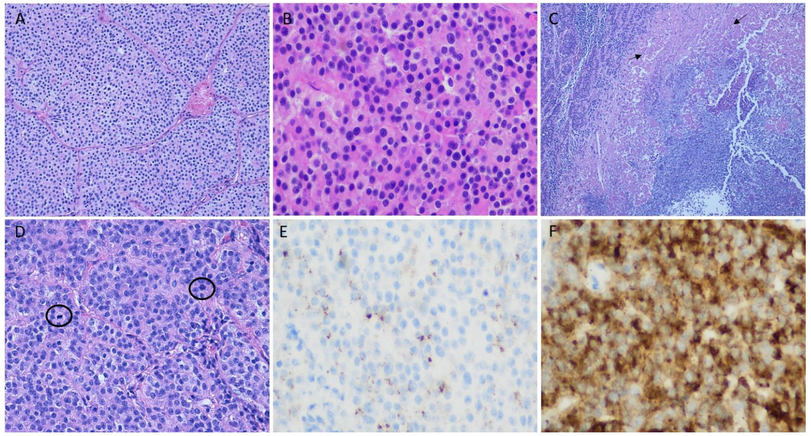

Figure 5. (A) H&E (200x) with areas of solid sheets separated by fibrovascular septae (B) H&E (400x) showing organoid arrangement of tumor cells with salt and pepper chromatin characteristic of a neuroendocrine tumor (C) H&E (200x), tumor cell necrosis (D) H&E (400x), highlighted mitotic activities (E) IHC, chromogranin, focally positive (F) IHC, PTH, strongly and diffusely positive in tumor cells

Lab	Normal	POD0	POD1	POD4	POD9	POD14	POD30
Ca	8.6-10.0 mg/dL	14.0	8.9	6.4	7.2	6.6	9.3
PTH	19-88 pg/mL	678	44	222	281	356	29
ALP	31-125 U/L	372	411	722	1232	1107	521

Table 1. Postoperative laboratory monitoring.

FURTHER MANAGEMENT

- After removal of the tumor, she developed symptomatic hypocalcemia, with nadir calcium on postoperative day 4 (Table 1).
- Calcium gluconate infusion, oral calcium, vitamin D, and calcitriol were used to maintain calcium within range.
- By postoperative day 30, PTH and Ca normalized on high doses of calcitriol, calcium carbonate, and vitamin D.

CONCLUSIONS

- This is a rare presentation of a well-differentiated pulmonary neuroendocrine tumor producing extremely high levels of PTH with resultant severe hyperparathyroidism, PTH-mediated bone changes, and pathological fracture complicated by hungry bone syndrome following resection of the source of PTH.
- To our knowledge, this is the first report of a pulmonary neuroendocrine tumor producing PTH, as most cases of neuroendocrine tumor-induced hypercalcemia are PTHrP-mediated.

REFERENCES

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