



### Presented By:



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### Rare case of parathyroid carcinoma

A 65-year-old man with multiple medical comorbidities, that included kidney stones and multiple gastric ulcers with perforations, was presented with hypercalcemia on his routine blood work during his visit with primary care physician. His calcium levels were 10.8 (1/2013), 10.6 (2/2013), and 10.4 (3/2013). Work up for primary hyperparathyroidism revealed elevated level of parathyroid hormone (PTH) of 100 to 109, vitamin D level of 50, creatinine level of 0.9.

He was referred to endocrinologist Svetlana Shifrin-Douglas, M.D. He was tested negative for MEN-1 syndrome. His bone density study showed T-score of -2.1 in the femoral neck consistent with osteopenia (3/2013). Parathyroid Sestamibi scan on March 21, 2013 showed persistent uptake consistent with a left inferior parathyroid adenoma (Figure 1).

**Past medical history:** Pancreatic insufficiency, inflammatory bowel syndrome, COPD, history of pneumothorax with a lobectomy in 1998, diverticulosis, septic shock in the past, cholecystectomy, subtotal gastrectomy at age 26 with G-tube placement (still has it). No family history of cancers or hyperparathyroidism. Physical examination was unremarkable, as well as neck examination and no neck lymphadenopathy. Neck ultrasound evaluation revealed the presence of a left inferior parathyroid adenoma (Figure 2).

He was diagnosed with primary hyperthyroidism complicated by nephrolithiasis, osteopenia and referred for surgery, parathyroidectomy, with endocrine surgeon Alexander Shifrin, M.D.

During surgery Dr. Alexander Shifrin found that his parathyroid gland appeared to be small but with obvious signs of invasion into the esophageal wall and surrounding tissue. Intraoperative pathology evaluation was consistent with parathyroid carcinoma. Immediate extensive resection was performed which included surrounding soft tissue with lymph nodes, partially esophageal muscle, and also left thyroid lobectomy was performed. Final pathology was consistent with parathyroid carcinoma involving left inferior parathyroid gland with invasion (Figure 3).

Follow up with Dr. Svetlana Shifrin-Douglas a few weeks after the surgery showed low normal calcium and PTH levels consistent with excellent outcome. A CT scan showed no evidence of recurrence. In two years after the surgery his Ca levels were between 8.8 and 9.2 and a PTH level of 28, PET/CT, CT scan and neck showed no evidence recurrence.



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### Rare case of parathyroid carcinoma

This patient had very unusual presentation of parathyroid carcinoma in the sense that his calcium and PTH levels were slightly above normal which is not typical for this disease.

Parathyroid carcinoma is a rare malignant neoplasm of the parathyroid gland that is affecting less than 1% of all patients with primary hyperparathyroidism. It may occur sporadically or as a part of a genetic syndrome. It has an indolent growth with a tendency for local invasion. Because of the rarity of the disease, lacking of definitive diagnostic markers, and clinical features mimicking benign primary hyperparathyroidism, preoperative diagnosis is difficult to establish preoperatively. As a result initial surgical treatment is inadequate essentially leading to disease recurrence where complete cure is unlikely. Parathyroid carcinoma should be suspected in the presence of markedly elevated PTH levels (>5 times the upper limit of normal) and serum calcium concentrations (above 14 to 15 mg/dL). The pathologic diagnosis of malignancy is challenging. Fine needle aspiration (FNA) biopsy of the parathyroid gland prior to initial operation is not recommended due to technical difficulty in differentiating benign and malignant disease on cytology specimens and the possible associated risk of tumor seeding from the needle track. Imaging techniques such as neck ultrasound and Sestamibi scan can help localize disease, but they are not specific for the cancer and not useful in the assessment of malignancy. PET CT is useful for the detection of metastatic disease.

**Treatment:** Complete, *en bloc* surgical resection of the tumor with clear margins remains the best chance of cure. Although prolonged survival is possible with recurrent or metastatic disease, cure is rarely achievable. Persistent or recurrent disease occurs as high as in 50% to 80% of patients with parathyroid carcinoma.

**Prognosis:** The reported survival rate is less than 50% to 85% at 10 years. Local recurrence, after surgery with *en bloc* resection, occurs at regional lymph nodes in 30% (between 22% and 60%) of cases, while distant metastases most frequently involve the lungs, liver, and bone.

Figure 1

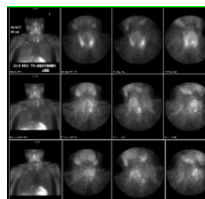
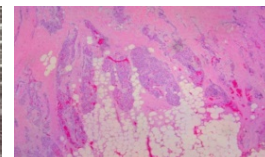


Figure 2



Figure 3



**Figure 1.** Sestamibi scan showing persistent uptake corresponding to the left lower parathyroid gland. **Figure 2.** Thyroid-parathyroid ultrasound showing enlarged left lower parathyroid gland. **Figure 3.** Pathological evaluation with H&E staining: parathyroid gland with area of invasion into fat and dense stromal fibrosis (50x)