Cause of Recurrent Hyperparathyroidism: Parathyromatosis, Parathyroid Cancer or Both

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Background

- Recurrent hyperparathyroidism is the reappearance of hypercalcemia after a normocalcemic period of at least 6 months post-parathyroidectomy, commonly due to incomplete removal of a parathyroid adenoma.
- Parathyromatosis is a rare cause of recurrent hyperparathyroidism where hyperfunctioning parathyroid tissue implants in the neck and mediastinum.
- Parathyroid carcinoma must be excluded histopathologically if parathyromatosis is suspected.
- Patients with parathyroid carcinoma usually have more markedly elevated calcium than typically seen in parathyromatosis.
- We present an interesting case of recurrent hyperparathyroidism secondary to parathyromatosis and parathyroid cancer.

Case Presentation

- A 50-year-old man with past medical history of stage 4 CKD and primary hyperparathyroidism presents with recurrence of hypercalcemia despite partial parathyroidectomy.
- Patient initially presented with weakness, nausea and vomiting.
- Vitals were consistent for BP 146/82, HR 90, RR 18, T 37.2
- Head and neck, cardiopulmonary, abdominal and lower extremity exams were normal
- Upon initial presentation patient had the following lab findings:

Laboratory Findings	
Calcium 13.3mg/dl (8-10mg/dl)	PTH 564pg/ml (10-15pg/ml)

The patient underwent left lower parathyroidectomy with pathology negative for malignancy.

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He remained normocalcemic for 2 years until he presented with weakness, nausea, and increased thirst.

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Calcium 15.2mg/dl	PTH 2485pg/ml

He was treated with IVF, calcitonin and cinacalcet with improvement in calcium levels to 10.2mg/dl

↓

Two months later, he presented with generalized weakness, tachycardia, and confusion.

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Calcium 17.3mg/dl	PTH >2500pg/ml

- A repeat parathyroid uptake scan revealed increased activity in the isthmus **(Figure 1)**
- The parathyroid tissue located near the isthmus was excised.
- Per intraoperative records, the parathyroid tissue was likely due to parathyroid implants from previous surgery.
- Pathology returned positive for malignancy**. (Figure 2 and 3)**

Images

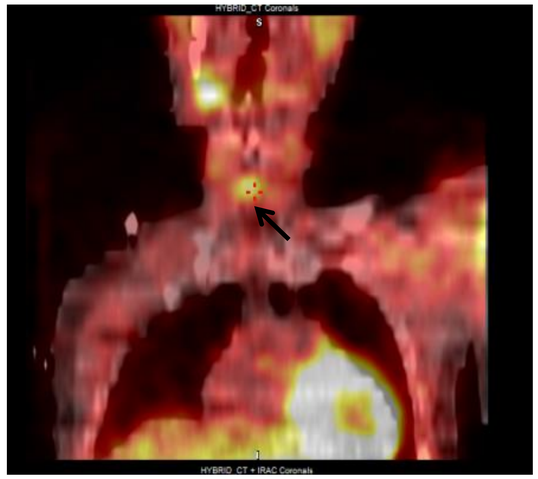

Figure 1. Delayed multiplanar SPECT images of the neck showing increased activity in the isthmus region with additional residual uptake over the left lower pole at the site of the initial surgery

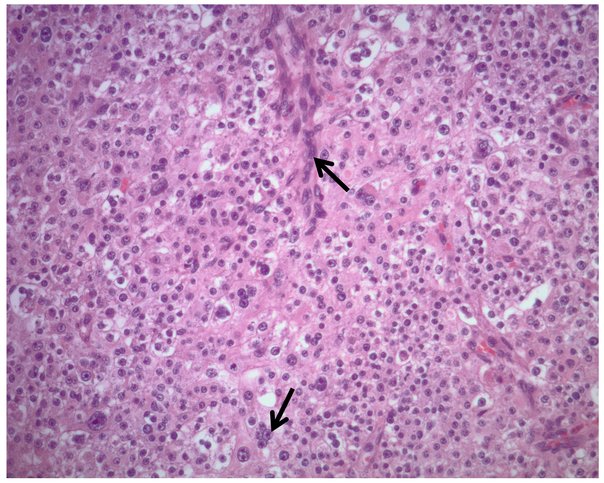

Figure 2. H&E histological slide showing parathyroid neoplastic cells consistent with parathyroid carcinoma

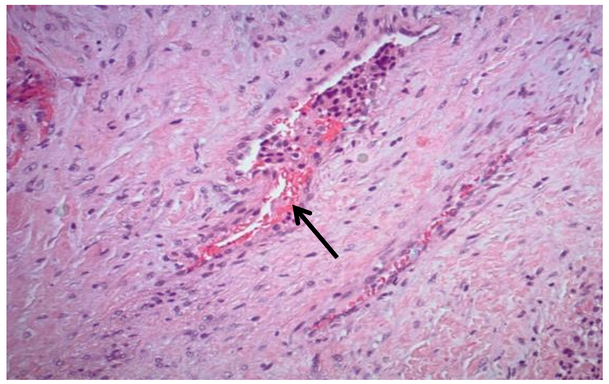

Figure 3. H&E histological slide showing parathyroid neoplastic cells consistent with parathyroid carcinoma. The parathyroid neoplasm is invading the soft tissue and shows vascular involvement.

Conclusion

- Parathyromatosis and parathyroid cancer are uncommon causes of recurrent hyperparathyroidism and should therefore be included in the differential diagnosis of recurrent hyperparathyroidism.
- Given our patient's early history of parathyroidectomy and most recent pathology positive for malignancy, he had both parathyromatosis and parathyroid cancer.
- In the the workup of recurrent hyperparathyroidism it is essential to consider parathyromatosis in addition to parathyroid cancer in the differential diagnosis.
- As in our patient's case, when he followed up in a month, his calcium level had improved but the PTH was still elevated. This was felt to be attributed to vitamin D supplementation as calcium normalized after discontinuation.

References

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2. Aksoy-Altinboga A, Sari A, Calli A. Parathyromatosis: Critical Diagnosis Regarding Surgery and Pathologic Evaluation. Korean J Pathol. 2012 Apr; 46(2): 197-200