Bilateral Water-Clear Cell Double Parathyroid Adenomas

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ABSTRACT

Objectives:
1) Describe the pathologic findings of water-clear cell hyperplasia and adenomas
2) Understand the clinical presentation of a patient with primary hyperparathyroidism caused by double water-clear cell parathyroid adenomas

Methods:
Case Report: A 66 year old Caucasian female was referred to our clinic with hypercalcemia, osteoporosis, and hyperparathyroidism. Sestamibi scan was non-localizing and thyroid ultrasound revealed bilateral thyroid nodules. She underwent total thyroidectomy and bilateral superior parathyroidectomy. The inferior parathyroid glands appeared grossly normal and were left intact.

Results:
Intraoperative PTH dropped from 209 to 39.5 and postoperative calciums returned to the normal range. Pathology revealed bilateral superior parathyroid adenomas with the right superior gland weighing 2.14 grams and the left weighing 1.27 grams. Both glands were consistent with water-clear cell hyperplasia. The thyroid gland, of which FNA had returned preoperatively as atypia, revealed multinodular goiter.

Conclusions:
WCC hyperplasia is a rare cause of primary hyperparathyroidism. To our knowledge only one other case has been reported that revealed bilateral water-clear cell parathyroid adenomas. Unlike primary chief cell hyperplasia, WCC hyperplasia is not associated with MEN syndromes and the incidence has decreased significantly over the past 25 years.

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INTRODUCTION

Primary hyperparathyroidism is most commonly the result of a single parathyroid adenoma in 87% to 91% of cases. The remaining culprits include four-gland hyperplasia, double adenomas, and rarely, parathyroid carcinoma. The reported incidence of double adenomas ranges from 2% to 15% of patients undergoing surgical therapy for primary hyperparathyroidism. The majority of parathyroid adenomas are of the chief or mixed cell type with relatively few of the oxyphil type identified. Water-clear cell adenoma of the parathyroid is a rare finding with only eight known cases previously identified and is composed of cells with vacuolated or granular cytoplasm. In contrast, water-clear cell hyperplasia is a well-documented cause of primary hyperparathyroidism but is also rare and the incidence has decreased over the past seventy-five years to less than 1% of cases of hyperparathyroidism.

CASE REPORT

A 66 year old Caucasian female was referred to our clinic with mild hypercalcemia, osteoporosis, and hyperparathyroidism. Sestamibi scan was non-localizing and thyroid ultrasound revealed bilateral thyroid nodules. Fine needle aspiration of the thyroid revealed atypia of the left thyroid nodule and the patient was also consented for thyroidectomy. She subsequently underwent total thyroidectomy and bilateral superior parathyroidectomy. The inferior parathyroid glands appeared grossly normal and were left intact due to the decrease of intraoperative PTH from 209 to 39.5 pg/mL. Postoperative calciums returned to the normal range and the patient did well postoperatively.

PATHOLOGY

The right and left parathyroid glands measured 2.5 and 2.0 and weighed 2.17 and 1.27 grams, respectively.
Both parathyroid glands showed histologically similar lesions composed of uniform, moderately sized sheets of clear cells with clear finely vacuolated cytoplasm (Figure 1).
The cells were present in nest and cords and separated by fine fibrovascular septa (Figure 2 and 3).
Distinct cell membranes were present and the nuclei show minimal atypia, with finely stippled chromatin (Figure 4).
The right parathyroid had a minute rim of extracapsular, histologically unremarkable parathyroid tissue (Figure 5).

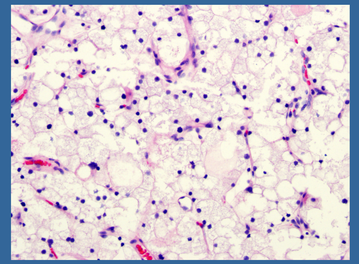

Figure 1

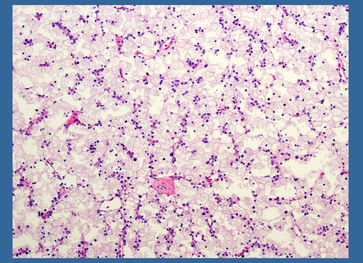

Figure 2

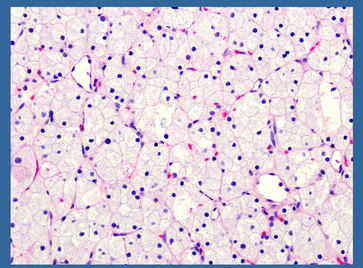

Figure 3

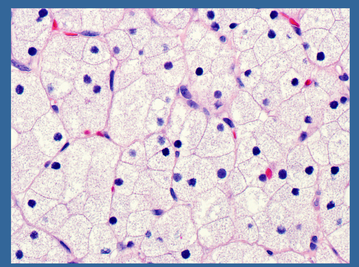

Figure 4

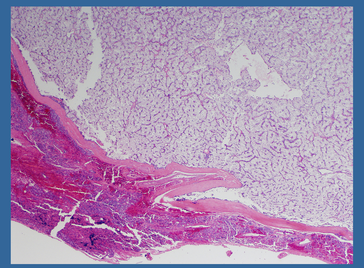

Figure 5

DISCUSSION

WCC hyperplasia is a rare, but well documented cause of primary hyperparathyroidism that has decreased to less than 1% of all patients undergoing surgical treatment of primary hyperparathyroidism. Interestingly, when first described by Albright in 1934, 12.8% of the 47 cases reported were consistent with WCC hyperplasia. Water-clear cell hyperplasia is characterized by proliferation of vacuolated water clear cells in multiple parathyroid glands and represents the only known parathyroid disorder in which the superior glands are larger than the inferior glands. Histologically, the appearance of water clear cell hyperplasia resembles renal cell carcinoma. Light microscopy reveals diffuse proliferations of clear cells characterized by clear cytoplasm and small dense nuclei, and higher magnification reveals cytoplasm filled with small vacuoles. To our knowledge only one other case has been reported that revealed bilateral water-clear cell parathyroid adenomas and total of eight case reports revealing WCC adenomas. Our case demonstrates the use of intraoperative PTH with subsequent decrease suggesting bilateral parathyroid adenomas which were confirmed histologically. Although the differentiation between parathyroid adenoma and hyperplasia is difficult, the use of intraoperative PTH is likely to result in adequate treatment of primary hyperparathyroidism defined by normocalcemia at 6 months after surgery.

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