An Undescended Intravagal Parathyroid Adenoma In A Patient With Multiple Adenomas and Supernumerary Parathyroid Glands

William R. Ryan, MD1 and John A. Ryan, MD, FACS2
1Department of Otolaryngology-Head and Neck Surgery, University of California-San Francisco, San Francisco, CA
2Department of Surgery, Virginia Mason Medical Center, Seattle, WA

ABSTRACT

A 22-year old woman presented with primary hyperparathyroidism to the Virginia Mason Clinic in Seattle, WA. Her serum calcium (Ca) and parathyroid hormone (PTH) levels were 11.7 mg and 121 units (with 80 being the upper limit of normal), respectively. A 24-hour urine test was 272 mg. She reported fatigue and bone and joint pain. She had no history of kidney stones, psychiatric disorders, abdominal symptoms, nor family history of hyperparathyroidism. An ultrasound showed no evidence of enlarged parathyroid glands. A sestamibi radioisotope scan showed no obvious focal signs, aside from a slight increase in uptake in the right central neck area.

During a right central neck exploration, a superior parathyroid adenoma and an inferior normal parathyroid gland (frozen section-confirmed normocellular) were found. With excision of the parathyroid adenoma, the PTH remained elevated. A left central neck exploration found a left superior parathyroid adenoma. After excision of the 2nd parathyroid adenoma, the PTH still remained elevated. Direct internal jugular vein sampling tests showed higher PTH levels on the left side (500, 1200, and 1811) compared to the right (188 and 194). The following steps were then performed: biopsies of candidate tissue in the left central compartment, left level 4 compartment, left upper mediastinum, and the right level 4 compartment. These frozen sections did not find any further parathyroid tissue. A left hemithyroidectomy was then performed. The dissection was discontinued. Postoperatively, the Ca and PTH levels remained elevated at 11.5 and 154, respectively.

Permanent section review confirmed the presence of a right superior and a left superior parathyroid adenoma and discovered another (third) parathyroid adenoma in the central thymic tissue that had been submitted only for permanent section.

Given the unilaterality of the venous sampling PTH elevations, a remaining undescended 4th parathyroid adenoma (5th parathyroid gland in total) was suspected in the left upper neck. Review of the original sestamibi scan found a possible area of slight increased uptake just superior and posterior to the left submandibular gland. A postoperative sestamibi showed more obvious uptake posterior to the left submandibular gland. A postoperative computed tomography scan of the neck with and without contrast demonstrated a mass located 4 cm superior to the left carotid bifurcation, in between the internal carotid and internal jugular vein (Figure 2).

A left upper neck exploration was performed within 2 months of her first operation. A 2 cm parathyroid adenoma was excised from the fibrous sheath of the vagus nerve (Figure 3). Postoperatively, the serum calcium and PTH were 9.3 and 16, respectively. Her vocal cord and swallowing function were both normal.

Two years postoperatively, her serum calcium and PTH have been normal. She has reported an increase in her energy level and a decrease in joint and bone pain.

Introduction

Parathyroid adenomas can be single or multiple and can be found in a range possible locations. Most parathyroid adenomas are single. Double adenomas are a well-known possibility up to even 9% of the time.1 Supranumerary parathyroid glands can occur up to 6.3% of the time.2 Parathyroid glands have been found anywhere from the parapharyngeal skullbase region to the middle mediastinum with specific locations that include: intrathyroidal, intrathyrmich within the carotid sheath, in the lateral neck, adjacent to the hyoid bone, and adjacent to the submandibular gland.3 Ectopic parathyroid glands and adenomas located superior to the thyroid gland appear to be the result of an incomplete descent of the 3rd brachial pouch during fetal development.4 Ectopic and supernumerary glands/multiple adenomas should be considered when managing patients with primary hyperparathyroidism. The following report is an unusual case of an undescended intravagal parathyroid adenoma in a patient with multiple adenomas and supernumerary parathyroid glands.

Case Report

A 22-year old woman presented with primary hyperparathyroidism to the Virginia Mason Clinic in Seattle, WA. Her serum calcium (Ca) and parathyroid hormone (PTH) levels were 11.7 mg and 121 units (with 80 being the upper limit of normal), respectively. A 24-hour urine test was 272 mg. She reported fatigue and bone and joint pain. She had no history of kidney stones, psychiatric disorders, abdominal symptoms, nor family history of hyperparathyroidism. An ultrasound showed no evidence of enlarged parathyroid glands. A sestamibi radioisotope scan showed no obvious focal signs, aside from a slight increase in uptake in the right central neck area.

During a right central neck exploration, a superior parathyroid adenoma and an inferior normal parathyroid gland (frozen section-confirmed normocellular) were found. With excision of the parathyroid adenoma, the PTH remained elevated. A left central neck exploration found a left superior parathyroid adenoma. After excision of the 2nd parathyroid adenoma, the PTH still remained elevated. Direct internal jugular vein sampling tests showed higher PTH levels on the left side (500, 1200, and 1811) compared to the right (188 and 194). The following steps were then performed: biopsies of candidate tissue in the left central compartment, left level 4 compartment, left upper mediastinum, and the right level 4 compartment. These frozen sections did not find any further parathyroid tissue. A left hemithyroidectomy was then performed. The dissection was discontinued. Postoperatively, the Ca and PTH levels remained elevated at 11.5 and 154, respectively.

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Discussion

The identification of a normal parathyroid in this case (right inferior) confounds the possibility of definite generalized parathyroid hyperplasia. Undescended parathyroid adenomas have been shown to account for 7% of persistent hyperparathyroidism in a large series as was the situation in this patient.7

Intravagal parathyroid adenomas are very rare having only been reported a few times in the literature.6-8 In these reports, the adenomas all represent supernumerary tissue similar to this case. One of these reports resulted in temporary vagal nerve dysfunction.

In patients with multiple abnormal glands, such as with this case, preoperative localizing tests are rarely able to identify all of the abnormal glands.5-10

Conclusion

This case demonstrates unusual possibilities in patients with primary hyperparathyroidism.

References