Undescended Superior Parathyroid: A Case Report

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Abstract

Background: Aberrant migration of parathyroid glands from their embryologic origin may result in undescended parathyroid glands. We present a case of an ectopic parathyroid adenoma at the level of the pyriform sinus. Methods: A 41-year-old female was evaluated for primary hyperparathyroidism. Following non-localizing ultrasound and planar sestamibi imaging, the patient underwent...

1. Introduction

Parathyroid glands normally descend from their embryologic origin in the pharyngeal pouches to their final locations in the central neck. However, aberrant migrations can lead to one or more parathyroid glands being in non-anatomic locations, including retroesophageal space, thymus, mediastinum, or within the thyroid gland. Rarely they fail to migrate, resulting in an undescended gland. Adenomas arising from these locations should be considered when initial preoperative imaging fails to identify a target adenoma or after unsuccessful surgery. Here we present a case of an ectopic parathyroid adenoma arising at the level of the pyriform sinus.

2. Case Report

A 41-year-old female presented to our institution for evaluation of primary hyperparathyroidism. She had a 7-year history of hypercalcemia, with levels up to 11.5 mg/dL (reference range: 8.7 - 10.2 mg/dL) and PTH levels up to 101 pg/mL (reference range: 10 – 73 pg/mL). She has no pertinent family history and is a former smoker (0.25 ppd, 5 pack-years). Initially, she underwent an ultrasound (US) of the neck and ⁹⁹ᵐTc-MIBI single photon emission computed tomography (SPECT) imaging, which failed to identify any suspicious lesions or localizing parathyroid adenomas. Subsequent SPECT/CT (Figure 1) and 4-D computed tomography (CT) (Figure 2) demonstrated evidence of a 1.3 cm lesion superior to the thyroid at the level of the pyriform sinus on the left side, likely representative of an ectopic or undescended parathyroid adenoma. Given its unusual location patient was preoperatively counseled on the risk of injury to the hypoglossal nerve, marginal mandibular nerve, and the possible need for a second incision for four gland exploration if the candidate lesion was not an adenoma.

On the day of surgery, pre-operative PTH level was 80 pg/mL. In the operating room, an upper transcervical incision was performed to obtain access to the identified site. The subplatysmal flap was raised superiorly and inferiorly to expose the inferior aspect of the submandibular gland. Then the investing fascia was incised just caudal to the submandibular gland, which was retracted superiorly to reveal the posterior belly of the digastric muscle. The digastric muscle was dissected along its anterior face and subsequently retracted superiorly to identify the hypoglossal nerve and the internal jugular vein. A crossing facial vein was encountered that required ligation to gain appropriate access. An enlarged and undescended parathyroid measuring 1.5 x 1.5 cm located deep and slightly inferior to the anterior belly of the digastric muscle, anterior and slightly medial to the carotid artery, was identified and carefully resected (Figure 3, Figure 4).
10 minutes following the excision of the adenoma, PTH fell to 16 pg/mL and at 15 minutes post-excision remained stable at 14 pg/mL indicating biochemical cure.

3. Discussion

Undescended parathyroid glands are typically defined as those located at or above the carotid bifurcation. This classification of parathyroid glands only constitutes 2-7% of parathyroid adenomas, making it the least common form of parathyroid glands found in ectopic locations. While rare, undescended parathyroid adenoma should be considered when initial preoperative imaging fails to identify a target adenoma or after unsuccessful parathyroid surgery.

Pre-operative localization studies are critical in the identification of possible undescended parathyroid adenomas. Imaging modalities typically include high resolution neck ultrasound, SPECT, parathyroid scintigraphy, CT and/or magnetic resonance imaging (MRI). In a meta-analysis of 1276 patients, Wong et al. demonstrate a sensitivity of 86% [Confidence Interval: 0.81-9.90] for $^{99m}$Tc-sestamibi SPECT/CT in identifying ectopic parathyroid adenomas, which is superior to the sensitivity of SPECT and planar imaging modalities alone.

The addition of anatomical imaging can further enhance the diagnostic localization of the candidate lesion. In a review of 656 patients at a single institution, Zerizer et al. found that combining $^{99m}$Tc-MIBI with anatomical scans (CT or MRI) significantly improved diagnostic accuracy (improving sensitivity and specificity to 100%). Anecdotally, the localization of parathyroid adenomas can be complicated by false positive signals generated by thyroid nodules as well as the salivary glands, as was the case for our undescended parathyroid. Additional imaging modalities, such as CT neck with contrast or MRI can be helpful to pinpoint the location and distinguish it from the neighboring structure as it did in this case. A recent review article demonstrated that MRI had both a sensitivity and specificity of up to 97% in the detection of parathyroid adenomas. Common MRI traits of adenomas can include elongated morphology, T2 fat saturation hyperintensity, and strong enhancement T1 post-contrast.

Four-dimensional CT has shown promise for the detection of ectopic parathyroid glands, but has the drawback of a greater radiation exposure compared to the sestamibi SPECT modality. Four-dimensional CT specifically used for localization of ectopic parathyroids has not been clearly defined in the literature, however, it demonstrates a higher sensitivity (82%) and specificity (92%) compared to other imaging modalities, suggesting its benefit as a useful adjunct to localizing ectopic parathyroid adenomas.

Importantly, in pre-operative planning, it is imperative to consider the potential need to perform a second incision in order to conduct a four-gland exploration in the case that the PTH did not decrease by > 50% after the removal of the adenoma. The unique location of the surgical incision employed for this patient lends itself to potential surgical complications that are otherwise uncommon in standard parathyroidectomies. Specifically, surgeons must consider the increased risk of injury to the hypoglossal and marginal mandibular nerves, and injuries to the carotid artery, internal jugular vein, and vagus nerve if dissection of the gland is required from these structures. Furthermore, the use of a two-incision surgical approach necessitates the surgeon to counsel patients on potentially managing two scars post-surgery. As such, it is important to note that pre-operative discussions with patients regarding surgical risks may differ from that of a standard parathyroidectomy.

4. References


5. FIGURES

![Figure 1](image-url)  
Figure 1. Sestamibi Single Photon Emission Computed Tomographic (SPECT/CT) Study. Yellow arrow points to the candidate lesion.
Figure 2. Preoperative 4D-CT scan. Yellow arrow points to the candidate lesion.

Figure 3. Removed ectopic parathyroid gland with the illustration of relevant anatomical structures.
Figure 4. Intraoperative image of surgical incision site.