ABSTRACT

The majority of primary hyperparathyroidism is caused by parathyroid adenomas and ectopic parathyroid adenomas are uncommon within this population.

Here we report a unique case of an ectopic parathyroid adenoma within the parapharyngeal space mimicking a carotid body tumor with congenital agenesis of the left common carotid artery. The diagnostic and technical challenges presented by this case highlight the importance of preoperative imaging and intraoperative parathyroid hormone assay in preventing unsuccessful identification of an ectopic parathyroid adenoma.

INTRODUCTION

The prevalence of ectopic parathyroid adenomas (PA) are low, representing 4-10% of all parathyroid adenomas [1]. Various locations of ectopic parathyroid glands have been described in the literature and an ectopic site within the carotid sheath remains rare.

Additionally, common carotid artery agenesis is an exceedingly rare congenital anomaly characterized by separate origins of the internal and external carotid arteries directly from the aortic arch [2]. Here we present a case of congenital agenesis of the common carotid artery associated with an ectopic PA within the parapharyngeal space mimicking a carotid body tumor.

This report also highlights the importance of preoperative imaging and the utility of intraoperative parathyroid hormone (PTH) measurement to positively identify an ectopic PA in the setting of aberrant carotid anatomy.

CASE REPORT

A 75 year-old gentleman with primary hyperparathyroidism (HPT) presented to the otolaryngology service in April 2011. His past medical history was significant for hypertension, type 2 diabetes mellitus, hyperlipidemia and prostate cancer. He denied any history of kidney stones, osteopenia or depression. There was no known history of thyroid carcinoma or pheochromocytoma except for parathyroid adenomas in the patient’s brother and sister.

Laboratory studies demonstrated a PTH value of 221 pg/mL and serum calcium of 12.2 mg/dL. Physical exam was unremarkable except for bilateral lower extremities burn wounds. There was no palpable cervical lymphadenopathy.

Initial 99mTc-sestamibi scintigraphy was performed and revealed asymmetry in the left neck region but did not localize the parathyroid adenoma (Figure 1). A contrasted neck CT scan demonstrated a 4.0 x 2.5 x 1.0 cm lesion in the left parapharyngeal space (Figure 2) suspicious for a paraganglioma based on its location, with no parathyroid adenoma identified.

DISCUSSION

80 to 85% of primary hyperparathyroidism (HPT) are due to parathyroid adenomas (PA). Ectopic PAs account for 4-10% of all PAs [1] and 20-25% of HPT [3] secondary to abnormal migration during embryogenesis. Inferior parathyroid glands are more prone to ectopic locations than the superior parathyroid glands due to a longer course of descent. The most common locations accounting for over 50% of all ectopic PAs are the thymus and anterior mediastinum [1]. Other unusual sites have been described in the literature but the incidence of an undescended parathyroid adenoma within the carotid sheath is extremely rare [4].

Ectopic parathyroid glands often present diagnostic and operative challenges and are common reasons of failed initial parathyroidectomy. Current practice of combined sonography and 99mTc-sestamibi can localize solitary adenomas with a sensitivity of as high as 95% [3]. Contrasted CT scan can provide additional anatomic information from the skull base to mediastinum in detecting most ectopic PAs that demonstrate intense enhancement [3]. It may aid in operative planning of patients with altered anatomy as in our case.

A carotid body tumor was initially suspected evidenced by its location and vascularity on CT neck and angiogram. However, the significant decrease of intraoperative PTH post-excision of the suspicious mass strongly supported the diagnosis of an ectopic PA, which was later confirmed by pathology. The intraoperative PTH monitoring did not only preclude cure but aided in the diagnosis of an ectopic PA in this particular case.

CONCLUSIONS

The successful identification and resection of the ectopic parathyroid adenoma presented here reiterate the importance of preoperative imaging studies that allow appropriate operative planning, as well as the utility of intraoperative parathyroid hormone assay in predicting cure during surgery.

Table 1

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<th>Pre-excision</th>
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<tr>
<td>Intra-operative PTH (pg/mL)</td>
<td>306</td>
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REFERENCES


Figure 1. 99mTc-sestamibi scintigraphy shows asymmetric tracer uptake in left submandibular region.

Figure 2. Axial, sagittal and coronal CT images of the neck demonstrate a left parapharyngeal space lesion.

Figure 3. Angiography image shows the absence of the left common carotid artery.

Figure 4. Operative image shows the relative location of the parathyroid adenoma.

Figure 5. Frozen sections of the parathyroid adenoma in H&E stain.

Postoperatively, the patient recovered well with a mild tongue deviation to the left and required calcium supplementation but remained asymptomatic for hypocalcemia at discharge on POD 6.

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