Mediastinal parathyroidectomy by VATS for persistent, sporadic primary hyperparathyroidism in an adolescent: a case report.

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Introduction

- Treatment for primary hyperparathyroidism (PHPT) includes surgical removal of the hypercellular parathyroid gland(s).
- ~20% of abnormal parathyroid glands are located ectopically (e.g., in the mediastinum) .
- Technetium-99 m ((99 m)Tc) sestamibi parathyroid scan with contrast-enhanced CT imaging is a first-line diagnostic study for tumor localization. Traditional CT imaging may be added to enhance anatomic precision, especially when lesions lie adjacent to large blood vessels.
- Depending on location, mediastinal parathyroid adenomas can be successfully removed via a traditional cervical surgical approach. However, many cannot be accessed with this approach. Alternative approaches include sternotomy or thoracotomy but the recent rise in minimally invasive techniques have been shown to reduce morbidity and improve postoperative recovery.
- Here, we report a case of a non-syndromic adolescent with persistent PHPT who was cured after undergoing mediastinal parathyroidectomy via VATS approach with intraoperative parathyroid hormone guidance and frozen-section pathologic analysis.

Figures

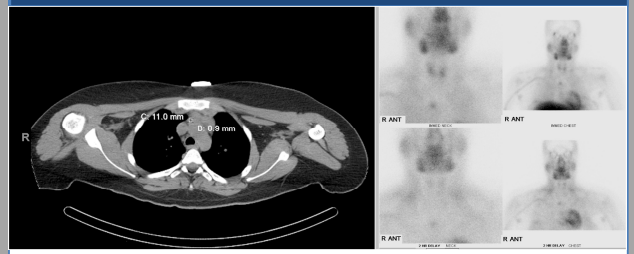


Fig. 1. Technetium-99 m ((99 m)Tc) sestamibi parathyroid scan showing mediastinal parathyroid adenoma adjacent to the proximal aortic arch.



Fig. 2. Computed tomography scan with contrast of the thorax showing mediastinal parathyroid adenoma adjacent to the aortic arch.

Case report

Preoperative work-up

- A previously healthy 13-year-old female presented to the Endocrine Surgery Clinic at the Penn State Health Milton S. Hershey Medical Center with recurrent renal stones and worsening fatigue. Her physical exam and family history were unremarkable.
- Labs: elevated serum calcium 11.7 mg/dL (reference: 8.4–10.2 mg/dL), elevated serum PTH 172 pg/ml (reference: 15–65 pg/ml), low vitamin D level 17 ng/ml (reference: 30–100 ng/ml), elevated 1,25-dihydroxyvitamin D level of 142 pg/ml (reference: 20–79 pg/ml), normal kidney function
- Surgeon-performed ultrasound of the neck suggested abnormal parathyroid glands in the tracheoesophageal groove bilaterally.
- Standard minimally-invasive neck exploration was performed. Three parathyroid glands seemed enlarged and were resected. Pathologic analysis confirmed hyperplastic parathyroid tissue in the specimens. However, serum PTH levels saw minimal decrease intraoperatively (dropped from 229 to 210 pg/ml).
- At two-week follow-up, the patient had persistent symptomatic hyperparathyroidism. A sestamibi parathyroid scan suggested a single ectopic parathyroid adenoma within the mediastinum (Fig. 1). Findings on thoracic CT scan with contrast were consistent (Fig. 2).

Operative approach

- Patient underwent surgical thoracic resection using a R-sided VATS approach after consultation with adult thoracic, pediatric cardiac and pediatric surgery faculty.
- Three ports were placed in the interspaces' medial to the anterior axillary line and CO2 insufflation was instituted with careful hemodynamic monitoring.
- A nodule suspicious for an ectopic parathyroid adenoma was present within fatty thymic tissue medial to the SVC and was removed. However, intraoperative serum PTH levels did not drop, and frozen-section analysis identified the nodule as lymphoid tissue.
- Resection of pericardial fat and the R thymus overlying the aortic arch based on preoperative imaging was then performed. A 0.7 × 0.5 × 0.3 cm pink-tan nodule was identified and isolated. The resected specimen was confirmed to be parathyroid tissue by frozen-section analysis and intraoperative serum PTH levels decreased from 210 to 24.2 pg/ml.

Postoperative course

- The patient recovered in the pediatric PACU and transferred to the pediatric hospital floor on the same day. She was started on 1500mg TID of calcium supplementation. She was discharged home on post-op day 2 with minimal discomfort and postoperative course was uneventful.
- At two-week follow-up, labs showed a normal PTH level of 47 pg/ml and total serum calcium of 7.8 mg/dL. Serial laboratory evaluation demonstrated normalization of her calcium level, with maintenance of a euhormonal state. She was able to wean off supplemental calcium.

Discussion

- PHPT in the pediatric population occurs in ~1 in 200,000 to 300,000 patients with most cases caused by a single adenoma [4]. Familial conditions such as MEN types 1 and 2a account for almost half of all cases of PHPT in children. PHPT secondary to an ectopic parathyroid adenoma such as in our young patient is rare [6].
- The most frequent cause of failed cervical exploration are ectopic parathyroid adenomas [1]. Ectopic parathyroid glands can be found in the cervical portion of the thymus, the posterior aspect of the middle third of the thyroid gland, the anterior mediastinum, the intrathyroidal region, or along the carotid sheath [7]. Ectopic or supernumerary parathyroid glands are more common in MEN patients than in sporadic hyperparathyroidism patients [8]. Our young patient with a single ectopic adenoma in the mediastinum had no evidence of familial disease.
- Precise localization of parathyroid glands is essential for complete surgical removal. Tc-99 m sestamibi scintigraphy is regarded as the best imaging modality to identify hyperfunctioning parathyroid tissue [9]. Neck ultrasound and 99mTc sestamibi scan are first-line imaging modalities; when combined with thoracic CT scan with contrast, diagnostic accuracy is improved [10]. In our case, a thoracic CT scan with contrast was able to increase resolution and further clarify the lesion's location and relationship to adjacent vessels.
- Our patient underwent a successful VATS procedure, following a cervical exploration that failed to achieve a euhormonal state. The VATS procedure has several advantages over median sternotomy or thoracotomy, including better visualization of tumor, less post-operative pain, reduced length of hospital stay, and superior cosmetic results [3]. Therefore, the VATS approach should be the preferred surgical method for removal of mediastinal parathyroid adenomas in adolescents. Rare complications include hemorrhage, subcutaneous emphysema, recurrent pneumothorax and pulmonary edema [11].
- This case report should raise awareness among clinicians of PHPT caused by ectopic parathyroid glands in adolescents even if they have no evidence of familial syndromes. More comprehensive imaging strategies should be employed prior to initial operation to reduce the rate of multiple surgical operations on a single patient.

Conclusion

In conclusion, ectopic mediastinal parathyroid adenomas are an uncommon cause of primary hyperparathyroidism in the pediatric population and the only curative option is surgical removal. We report a case of a pediatric, non-syndromic patient with anterior mediastinal and intrathymic parathyroid adenoma that was successfully localized via nuclear medicine and thoracic CT scans, then excised via VATS thymectomy, without complication and with resolution of elevated serum calcium and PTH.

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