Rare case of a hyperfunctional parathyroid cyst in a pediatric patient

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Background

Parathyroid cysts are rarely encountered, representing an estimated <1% of neck masses and 0.5-1% of parathyroid lesions[1]. The largest available case review describes a total of 359 cases from 1905 to 2016.[2] Parathyroid cysts are generally described as functional or nonfunctional based on their secretion of parathyroid hormone.

Functional parathyroid cysts account for approximately 10-33% of parathyroid cysts and are often discovered upon workup for symptomatic hypercalcemia.[3] It is estimated 1% of cases of primary hyperparathyroidism are caused by a functional parathyroid cyst. Functional parathyroid cysts have been encountered in a variety of locations from the angle of the mandible to the mediastinum.[2]

Nonfunctional parathyroid cysts are more common and often discovered incidentally. They are most often seen adjacent to the left inferior thyroid lobe. Both nonfunctional and functional parathyroid cysts can cause compressive symptoms including dysphagia, airway compression, and recurrent laryngeal nerve palsy.

These lesions are most often observed in patients age 30 – 60, with functional lesions more common in older patients within this range. Parathyroid cysts are seldom encountered in children, with 6 reported cases noted. The youngest age reported is 8. Available case reports describe nonfunctional cysts.[2,4–6]

Case Description

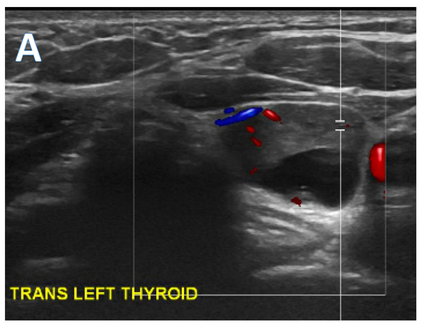
A 14 year-old female presents to otolaryngology clinic upon referral from endocrinology for symptomatic primary hyperparathyroidism. She was first diagnosed with renal stones at age 12. Since then, she has had recurrent stones despite conservative treatment and hydration. Her other symptoms included fatigue, bone pain, abdominal pain and constipation. Parathyroid hormone (PTH) and calcium (Ca) levels which were found to be elevated at 953 and 12.4 respectively. Prior to referral for surgical management, patient required admission for intravenous hydration and medical treatment of hypercalcemia with calcitonin, steroids, and cinacalcet.

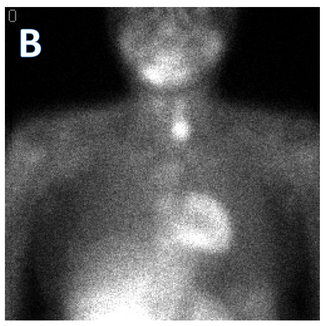
A thyroid ultrasound was performed and noted a 0.6 x 1.4 x 1.0 cm cystic lesion within the posterioinferior left thyroid gland. (Figure A) A distinct parathyroid lesion was not identified. Sestamibi scan and single-photon emission computerized tomography (SPECT) computed tomography showed radiotracer accumulation in this area concerning for parathyroid adenoma. (Figures B, C, D)

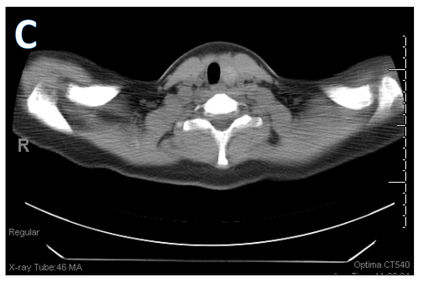
Due to concern for a thyroid cyst near the suspected adenoma, parents were consented for hemithyroidectomy as well as four gland exploration. The patient was taken to the operating room for parathyroidectomy. Initial intraoperative PTH level was 1,721. A midline approach was taken to the left inferior thyroid lobe, where a dark brown 1.5cm cystic lesion was encountered. A plane was encountered between the cystic lesion and the thyroid gland. Tissue with appearance more consistent with parathyroid gland was attached to the cyst posteriorly. The mass was removed en bloc and sent for frozen pathology. (Figure E)

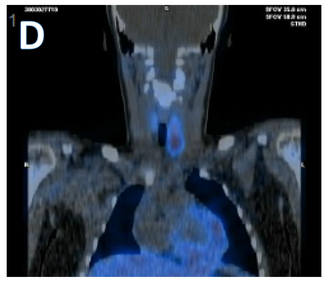
Lab Values

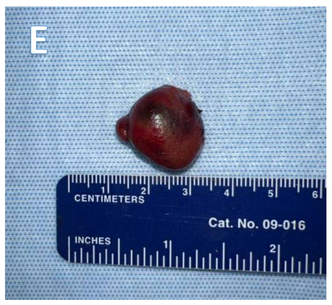
Time	Calcium (mg/dL)	PTH (pg/mL)
Pre-operative	12.4	953
Intra-operative manipulation		1721.4
Intra-operative 10 min		214.7
Intra-operative 30 min		120.7
PACU	9.0	58.0
POD 1	7.4	19.7
POD 4	9.4	54.9











Figures

A. Thyroid ultrasound demonstrating 0.6 x 1.4 x 1.0 cm cystic lesion

B. Tc99m-Sestamibi scan with asymmetric increased radiotracer simulation inferior to the left thyroid lobe

C. Axial non contrasted CT image of the neck demonstrating cystic mass inferior to left thyroid lobe.

D. Coronal SPECT-CT with increased radiotracer simulation at left inferior thyroid area corresponding with location of cystic lesion

E. Intraoperative specimen photo of parathyroid cyst

Case Description

PTH levels at 10 minutes and 30 minutes post-excision were 214 and 120. Frozen pathologic examination of the specimen confirmed the presence of parathyroid tissue. The case was concluded and the patient admitted for post operative monitoring of PTH and Ca levels. Patient was discharged on post operative day 4 once calcium levels stabilized on oral supplementation. Outpatient post operative course was complicated by hungry bone syndrome requiring prolonged calcium, calcitriol, sodium phosphate and potassium phosphate supplementation. At 6 months post op patient was off all supplementation without recurrence of kidney stones. Final pathology confirmed a 2.7g cystic parathyroid adenoma.

Discussion

Here we describe a case of a hyperfunctional parathyroid cyst in a pediatric patient. The diagnosis with made intraoperatively, with preoperative imaging suggesting presence of a cystic thyroid lesion. This finding was unexpected, as hyperfunctional parathyroid cysts are an uncommon cause of primary hyperparathyroidism in adults. No case report describing a functional parathyroid cyst in a pediatric patient has been published.

Diagnosis of parathyroid cysts is often made intraoperatively as ultrasound is not useful in distinguishing between thyroid or parathyroid origin of cysts. Sestamibi and SPECT imaging has also been shown to be less reliable for localizing hyperfunctional parathyroid cysts, with 29% of functional parathyroid cysts definitively localized on preoperative imaging compared to 68-95% of non-cystic adenomas.[7]

Preoperative fine needle aspiration may be used for diagnosis of parathyroid cysts. For non-fuctional cysts, aspiration of cyst contents can be both diagnostic and therapeutic. Cyst contents will have elevated PTH levels, however these levels do not necessarily specify if the cyst is functional or nonfunctional. Recurrent cysts have been successfully treated with ethanol sclerotherapy.[8] For functional parathyroid cysts, surgery is the treatment of choice. For patients who are not interested in pursuing surgery or who are poor surgical candidates, medical treatment with cinacalcet is an option.

Several theories exist regarding the origin of parathyroid cysts. These theories include origin from residual Kürsteiner's canal on the third branchial cleft, retention cyst of parathyroid secretions, coalescence of microcysts, or cystic degeneration of existing adenoma.[9] Theoretically functional cysts would be more likely due to cystic degeneration of an adenoma and nonfunctional cysts of congenital origin. Given the congenital theories, it is interesting that these lesions are seldom seen in pediatric populations.

Conclusion

Hyperfunctional parathyroid cysts have not been reported in children but must be considered in pediatric parathyroid surgery and patients with primary hyperparathyroidism and a cystic central neck nodule.

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