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Paediatric Cystic Parathyroid Adenoma

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Abstract

Presentation
A 15-year-old boy was admitted with a 4-week history of nonbilious, non-bloody emesis 3-4 times a day with progressively decreasing appetite with nausea, fatigue, irritability and 8 kg weight loss. His labs were normal except for BUN 12.1, Creatinine 150, Calcium 4.56 and PTH 157.9.

Diagnosis
Data was collected from the patient's medical chart in addition to radiology and histology reports.

Treatment
Mixed solid and cystic adenoma was treated with a parathyroidectomy.

Conclusion
Cystic parathyroid adenoma is an important consideration for individuals presenting with unexplained electrolyte imbalance, hyperparathyroidism and hypercalcemia.

Introduction

Cystic parathyroid adenomas are very rare and account for less than 1/10,000 of neck masses.[1] We report a 15-year-old patient presenting with hyperparathyroidism and hypercalcemia secondary to cystic parathyroid adenoma.

Case Report

A 15-year-old boy was admitted with a 4-week history of nonbilious, non-bloody emesis 3-4 times a day with progressively decreasing appetite with nausea, fatigue, irritability and 8 kg weight loss. His labs were normal except for BUN 12.1, Creatinine 150, Calcium 4.56 and PTH 157.9.

Apart from Attention Deficient Hyperactivity Disorder, his medical, surgical, family, social history, and systems review were unremarkable. He also had no allergies and his immunizations were up to date.

On examination, there was no evidence of cervical lymphadenopathy or a thyroid mass. The thyroid gland was palpable and non-tender. His cardiovascular, respiratory and gastrointestinal examinations were unremarkable.

The working diagnosis included parathyroid adenoma followed by parathyroid hyperplasia. Parathyroid carcinoma is less likely as it is extremely rare in the paediatric age group

On ultrasound, two cystic lesions were noted adjacent to the lower pole of the right thyroid lobe, measuring 2.3 x 2.0 x 3.2 cm and 2.9 x 3.5 x 5.2 cm. The smaller cyst appeared simple with a thin internal septation without internal vascularity, while the larger one appeared complex and demonstrated internal vascularity (Figure 1).

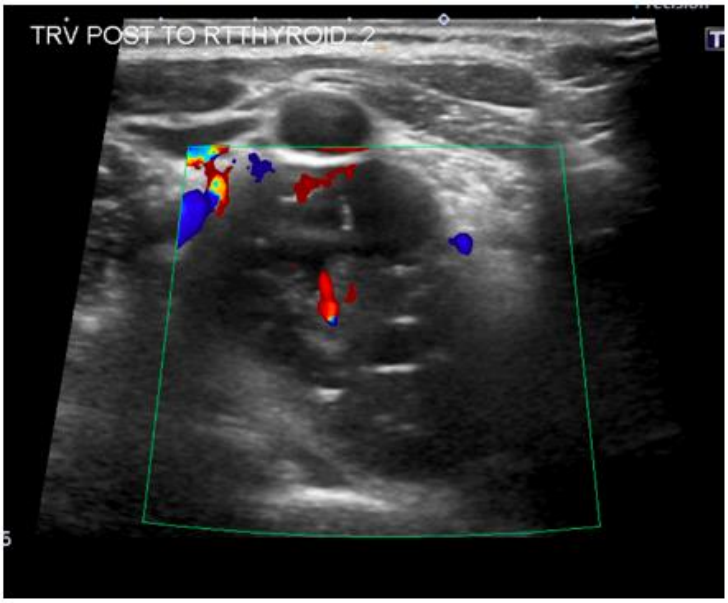

Figure 1: Ultrasound images of the neck at the level of the thyroid gland demonstrates vascular solid and cystic components within the lesion situated posteriorly to the carotid artery.

Parathyroid scintigraphy (^{99m}Tc- Sestamibi) revealed a focal increased tracer uptake at the inferior thyroid bed, right of the midline, which was further confirmed with SPECT/CT. When correlated with the neck ultrasound, these findings were consistent with a parathyroid adenoma.

Subsequently, the patient underwent a right-sided parathyroidectomy. The excised mass weighed 60.0 g and measured 8.0 x 5.0 x 3.6 cm (Figure 2). Histological examination demonstrated a biphasic lesion with about 30% being a pale solid irregular area while the other 70% was cystic and multilocular with three ill-defined locules. No features associated with malignancy were present apart from capsular invasion without extension to surrounding tissue and broad intra-tumoral fibrous bands containing expansile nodules. These findings were reflective of an atypical parathyroid adenoma.

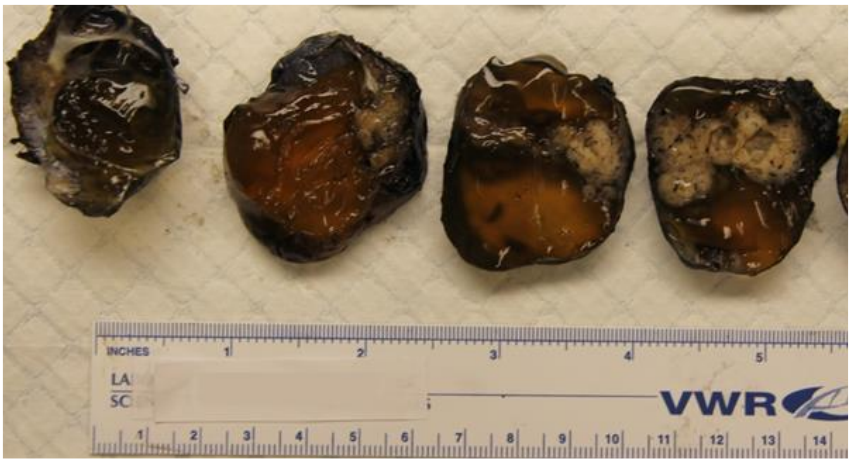

Figure 2: Sectioned Excised Mass (60g) demonstrating the nodular configuration with solid and cystic components. No features associated with malignancy were present apart from capsular invasion without extension to surrounding tissue and broad intratumoral fibrous bands containing expansile nodules. Additionally, there was no evidence of vascular or perineural invasion, and no metastasis to surrounding tissues. Findings were most in keeping with atypical parathyroid adenoma, which is a diagnostic term applied to a parathyroid neoplasm exhibiting some atypical features, but not fulfilling the absolute criteria for malignancy.

He was discharged and scheduled for follow up with Endocrinologyas an out-patient for his elevated parathyroid hormone levels (23.7 mmol/L). He remained asymptomatic at ten weeks postoperatively.

Discussion

Majority of parathyroid adenomas are solid; however, they can be cystic or partly cystic in 1-2% of Primary Hyperparathyroidism cases. In 1952, Green et al. reported the first case of a functioning parathyroid cyst causing primary hyperparathyroidism[4], and only around 300 cases of cystic parathyroid adenomas have been reported in literature thus far.[3, 4] These lesions are more common in males between the ages of 40 and 50 years involving predominantly the inferior parathyroid glands[5] and complex parathyroid adenomas are very rare in children. Multiple explanations have been put forward regarding the pathogenesis of cystic parathyroid adenomas, including (i) vestigial origin and development from the 3rd and 4th branchial cleft and cyst, (ii) amalgamation of parathyroid acini, (iii) parathyroid development failure, (iv) retention of secretion vesicles, and (v) intra-adenoma hemorrhage with consecutive liquefaction of hematoma.[5]

Diagnosing parathyroid adenomas preoperatively poses many challenges due to cytologic similarities in parathyroid cysts and non-functional thyroid cysts along with the low sensitivities of ^{99m}Tc-Sestamibi and neck ultrasound for cystic adenomas compared to solid adenomas. Consequently, neck ultrasound results should be correlated with ^{99m}Tc-Sestamibi to increase diagnostic sensitivity for cystic adenomas.

Cystic parathyroid adenomas are a subtype of atypical parathyroid adenomas. These can present in the paediatric population, although they are much more common in males between the age of 40-50 years old. Additionally, atypical parathyroid adenomas share some similarities with parathyroid carcinomas, and therefore an accurate diagnosis is essential for timely and effective management. Consequently, cystic parathyroid adenomas should be on the differential diagnosis list for individuals presenting with unexplained electrolyte imbalance, hyperparathyroidism and hypercalcemia.

Declaration of Conflicts of Interest:
No conflicts of interest to declare.

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