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Journal

Oxford Medical Case Reports, 2019(12)

ISSN

2053-8855

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Publication Date

2019-12-31

DOI

10.1093/omcr/omz129

Peer reviewed

OMCR

Oxford Medical Case Reports, 2019;12,519–523

doi: 10.1093/omcr/omz129
Advance Access Publication Date:
Case Report

CASE REPORT

Undescended retropharyngeal parathyroid adenoma with adjacent thymic tissue in a 13-year-old boy with primary hyperparathyroidism

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Abstract

We describe a rare presentation of a symptomatic parathyroid adenoma located in an ectopic retropharyngeal position in a 13-year-old boy. Preoperative CT scan and MRI demonstrated the ectopic location of the parathyroid adenoma. The patient underwent successful parathyroidectomy with cure of his hyperparathyroidism. On pathologic exam, the specimen was made up of a parathyroid adenoma and adjacent thymic tissue, indicating that it was likely an undescended lower parathyroid gland arising from the third pharyngeal pouch. Ectopic retropharyngeal parathyroid adenomas are very rare and to our knowledge, none have been previously described in adolescents.

INTRODUCTION

Primary hyperparathyroidism has an estimated incidence of 1 per 200 000–300 000 and a prevalence of 2–5 in 100 000 in the pediatric population [1]. In adolescents, approximately 80–92% of primary hyperparathyroidism is due to a single parathyroid adenoma [2]. In 80–85% of these patients, the adenomas are located in typical anatomical locations, while in 15–20% they are ectopic [3]. Ectopic parathyroid adenomas can be located at any location along the embryologic pathway of descent, with the most common locations being the anterior mediastinum, thymus, retroesophageal region or intrathyroidal [4]. The retropharyngeal location is rarely reported for parathyroid adenomas [5–9]. The retropharyngeal location of the parathyroid gland is thought to be due to the common embryologic origin of the superior parathyroid gland and the apex of the piriform sinus from the fourth branchial pouch. If the gland fails to separate from the piriform sinus, the structures may migrate together to a retropharyngeal location [9].

CASE REPORT

The patient being presented is a 13-year-old African-American male that presented to the emergency department with a

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Received: July 4, 2019. **Revised:** October 3, 2019. **Accepted:** November 7, 2019

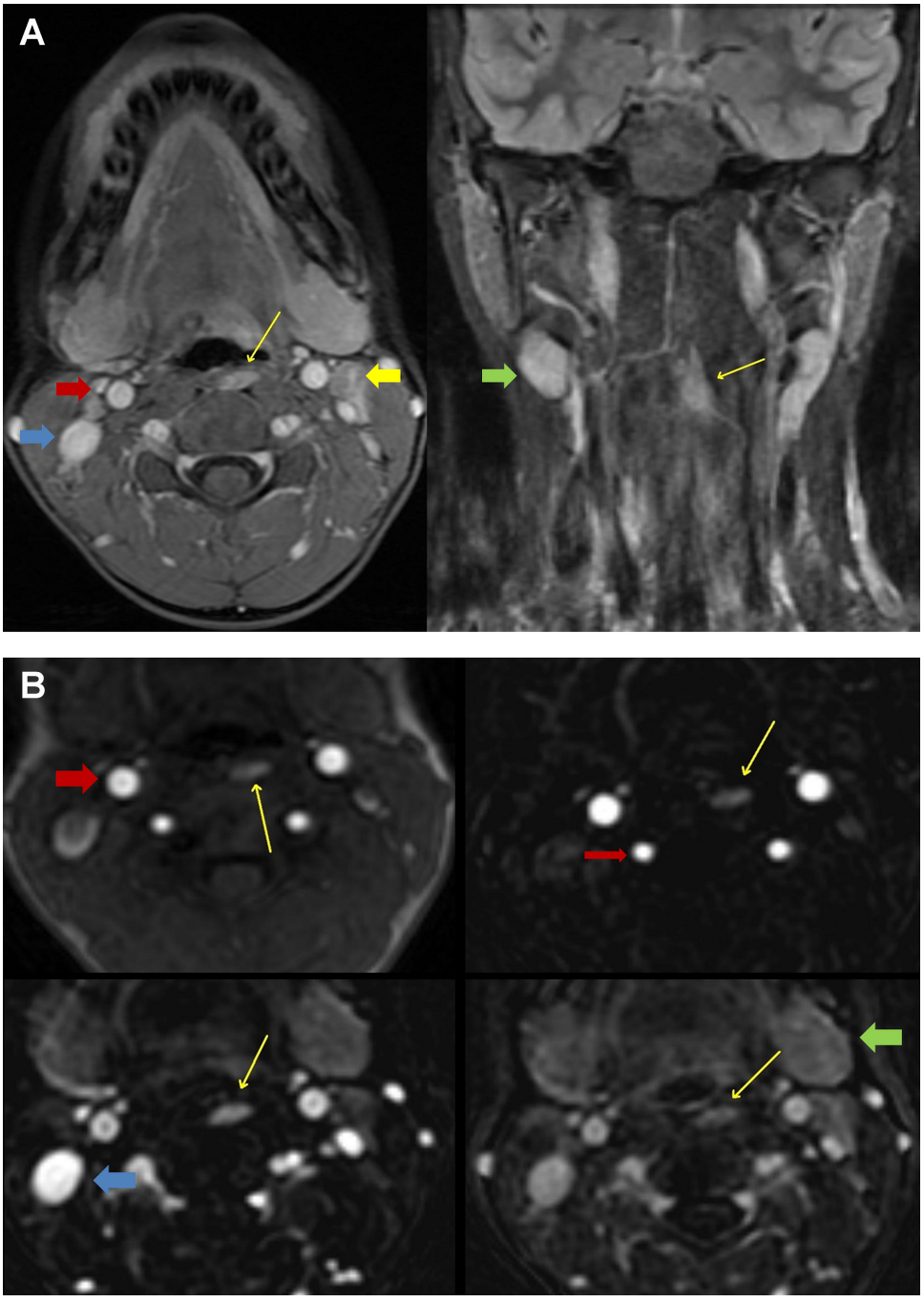

Figure 1: MRI with and without contrast, and perfusion imaging: (A) Axial T1 Post-contrast (left; TR: 7.8 ms, TE: 3.7 ms) and Coronal STIR (right; TR: 3050 ms, TE: 43 ms) demonstrating an oblong, enhancing area (narrow yellow arrows) in the left prevertebral soft tissues just cranial, posterior and medial to the upper pole of the left lobe of the thyroid gland (not visible on these sections of interest), that did not have MRI characteristics of vascular (red arrow = internal carotid artery; blue arrow = internal jugular vein) structures or lymph nodes (thick yellow arrow). Submandibular gland is indicated with a green arrow. (B) Magnetic Resonance Perfusion images, area of interest annotated with arrows. Top left to bottom right: early arterial (unsubtracted); early arterial, arteriovenous and late arteriovenous (subtracted) images demonstrating the enhancing area in the left prevertebral tissues that does not wash out on late perfusion images, suspicious for a parathyroid adenoma. (thick red arrow = internal carotid artery; narrow red arrow = vertebral artery; blue arrow = internal jugular vein). Submandibular gland is indicated with a green arrow. Thyroid gland is not visible on these planes of interest.

3 day history of fever and sore throat with associated difficulty swallowing. He was diagnosed with right peritonsillar cellulitis and underwent bedside incision and drainage. In the course of his workup, labs included a metabolic panel that showed that his kidney function was normal with a serum creatinine of 0.93 mg/dl (rr 0.4–1.00) and BUN of 17 mg/dl (rr 7–18) while having hypercalcemia with calcium of 14.7 mg/dl (rr 8.8–10.8), and ionized calcium elevated at 1.68 mmol/L (rr 1.1–1.32).

After hydration his calcium dropped to 12.6 mg/dl, and his iPTH was elevated at 124.8 pg/ml, urinary calcium was 23.2 mg/dl

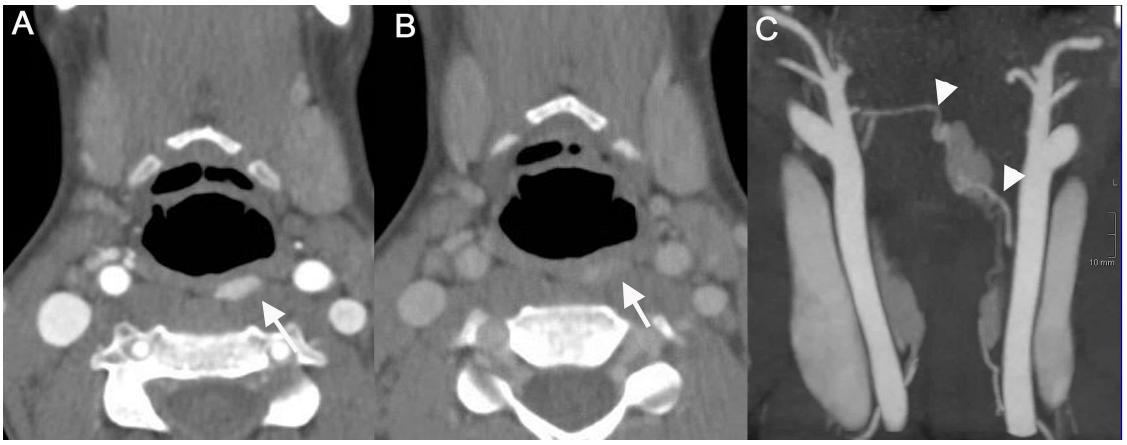

Figure 2: Contrast-enhanced CT imaging was performed with a limited field of view of the area of suspected midline retropharyngeal parathyroid adenoma with arterial angiographic and 40-s delayed venous phases confirming arterial phase enhancement of 293 Hounsfield units (A, arrow) and rapid wash-out to 127 Hounsfield units (B, arrow). Coronal reconstructions from the 0.625 mm source axial arterial phase sequence better defined the bilateral vascular structures (C, arrowheads) for surgical planning.

and urinary creatinine was 44.8 mg/dl, giving a urinary calcium:creatinine ratio of 0.52, which is elevated (normal is <0.21) reflecting hypercalciuria. His 25OHD was low at 13 ng/ml (considered vitamin D deficient when <20 ng/ml). The hyperparathyroidism was not secondary to the vitamin D deficiency, as it was associated with severe hypercalcemia. He did not have an elevated IGF1, prolactin or a fasting gastrin to suggest MEN type 1. His calcitonin was suppressed. The patient did not have problems with nocturia or polyuria. The family denied any recent constipation, although he previously had issues with chronic constipation. They did not note any behavioral changes or irritability or abdominal pain. However, the family did note that the patient was not as active as other children his age, which had been present for years. They stated that he walked as if his bones hurt. Participation in physical education at school was difficult for him. There was no known family history of kidney stones, pituitary problems, or galactorrhea.

Initial nuclear medicine parathyroid sestamibi scan was read as within normal limits. A multiplanar multisequence magnetic resonance imaging (MRI) of the neck/face without and with intravenous gadolinium contrast was obtained. Additional dynamic 4D contrast-enhanced magnetic resonance (MR) perfusion imaging was performed; subtraction sequences were generated, which suggested a possible ectopic parathyroid adenoma in the left retropharyngeal soft tissues, just cranial, posterior and medial to the upper pole of the left thyroid lobe, but not definitive (Fig. 1). The oblong, enhancing focus appeared to wash-in and does not wash out on the late perfusion images. Subjective peak enhancement also appeared to be more than cervical lymph nodes. Upon further discussion with the radiologists, they stated that based on the unusual location the imaging was not entirely definitive. He subsequently underwent a parathyroid CT protocol consisting of arterial (CT Angiographic) phase and delayed (45 s after completion of the 1st phase). Field of view was constrained from C2-C7 to reduce dose, given the pre-existing MRI findings excluding other ectopic lesions. The CT scan suggested a 1.3 cm ectopic retropharyngeal parathyroid adenoma (Fig. 2).

The patient underwent parathyroidectomy through a high transverse cervical incision. The mass was located in the left retropharyngeal space very close to the anterior portion of the cervical spine. The specimen was approximately 3 cm in length and sent to pathology for examination (Fig. 3). The tissue pathology revealed an enlarged parathyroid gland, which was cellular and surrounded by a thin, fibrous capsule. The parathyroid tissue was composed of one type of cells arranged in a vague, organoid pattern, with many cells showing enlarged nuclei. In addition to the parathyroid tissue, a large portion of the specimen was thymic tissue with scattered Hassall's corpuscles (Fig. 4).

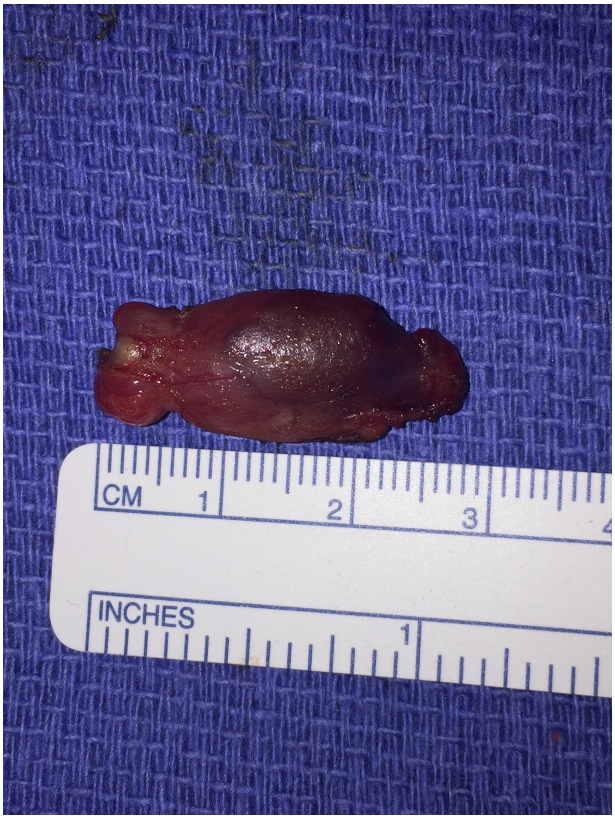

Figure 3: Ectopic left retropharyngeal parathyroid adenoma measuring 3.0 cm.

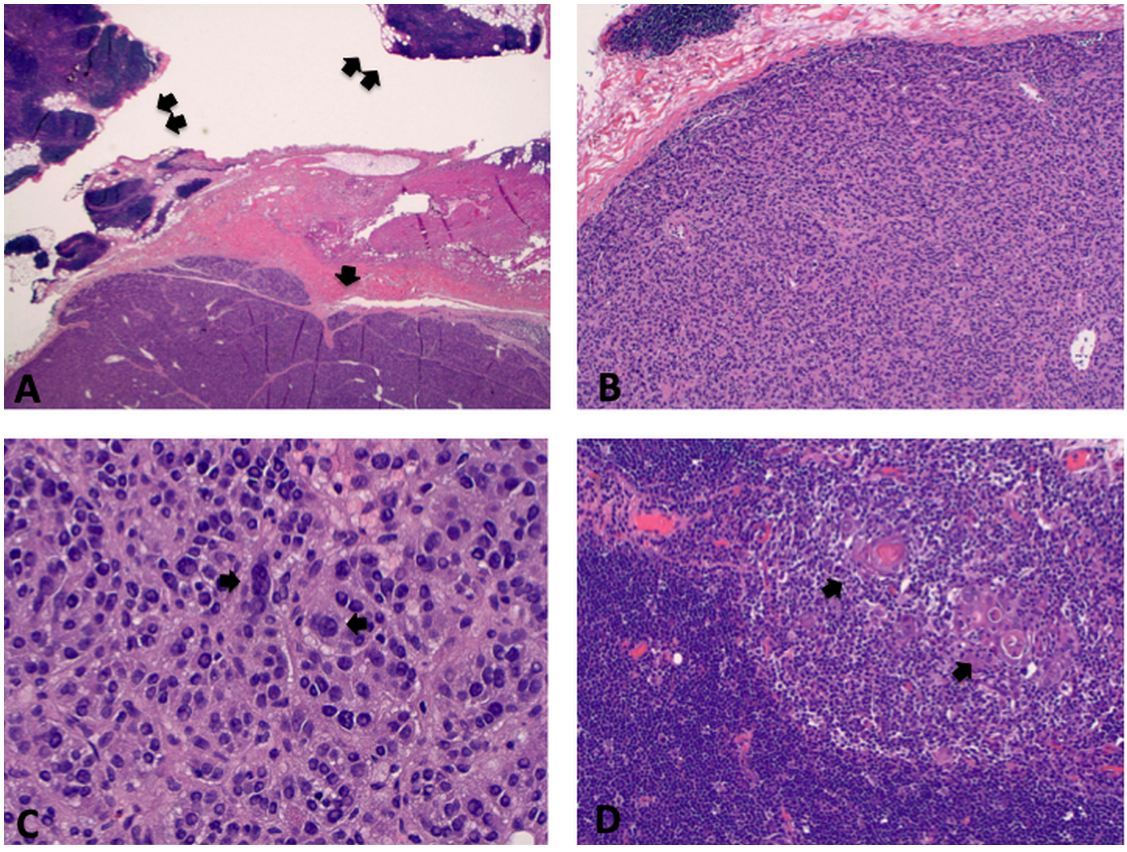

Figure 4: Low-power view of parathyroid adenoma (one arrow) and ectopic thymic tissue (two arrows) (A, H&E, ×10). Parathyroid adenoma-encapsulated and circumscribed cellular nodule (B, H&E, ×100) composed of one type of cells arranged in a vague, organoid pattern with few large bizarre nuclei (arrow) in high-power view (C, H&E, ×400). Normal-appearing thymic tissue with Hassall corpuscles (arrow) (D, H&E, ×100).

His intact PTH intraoperatively was 206.2 pg/ml (rr 11–74) and dropped to 39 pg/ml after the removal of the adenoma. About 90 min later the serum calcium was 11 mg/dl with a transiently low PTH of 5.2 pg/ml. By the next morning his calcium dropped slightly to 8.5 mg/dl as PTH was recovering to 12.1 pg/ml, and low dose calcium carbonate supplementation of 500 mg (200 mg elemental calcium) twice daily was added temporarily. On the second day after his surgery his PTH rose further to 22.8 pg/ml, and calcium carbonate was stopped the next day. Eleven days after his surgery, off any calcium supplementation, he had a normal serum calcium of 9.4 mg/dl and normal serum intact PTH of 32 pg/ml. As his PTH and calcium levels normalized, his symptoms related to hyperparathyroidism improved, being able to go back to full activity without feeling pain.

DISCUSSION

The superior parathyroid glands originate from the fourth pharyngeal pouch and are relatively constant in their location. In contrast, the location of the inferior parathyroid glands is extremely variable. They are derived from the third branchial pouch along with the thymus. They descend in the neck, crossing the path of the superior glands, and eventually lie in a position anteroinferior to the thyroid lobe or within the thyrothymic tract [10]. The thymus embryogenically arises from the paired third branchial pouches in humans and migrates along the course of the carotid sheaths into the anterior mediastinum where the two lobes fuse to form one gland. The failure of complete descent results in ectopic thymus or ectopic thymic tissue. Ectopic thymic tissue can be found at any level of the pathway of normal thymic descent from the angle of mandible to the superior mediastinum.

Most often, ectopic parathyroid adenomas are found in the upper mediastinum or posterior to the esophagus; however, there have been reports of parathyroid adenomas in other areas of the head and neck such as the pyriform sinus [11]. The patient presented in our case report had an extremely rare presentation of ectopic parathyroid adenoma with adjacent thymic tissue found in the retropharyngeal space. Although our patient underwent the traditional transcervical approach to parathyroidectomy, there have been several reports of transoral robotic approach to these tumors [5, 6].

Our case is also unique in that the patient was only 13 years old. Primary hyperparathyroidism affects ~1% of the population, occurring mainly in individuals aged 50–60 years. However, primary hyperparathyroidism in adolescents is rare. A recent review of the literature revealed less than ten cases of primary hyperparathyroidism due to ectopic adenomas being reported mainly in the mediastinum [12]. To our knowledge, our case is the first report of an ectopic retropharyngeal adenoma in an adolescent.

It must be noted that in this patient that the localization of the ectopic parathyroid adenoma on imaging, including dynamic 4D MRI, and confirmatory CT, was very important in guiding

the surgical approach and enabling the surgeon to successfully locate and remove the ectopic parathyroid adenoma.

CONFLICT OF INTEREST STATEMENT

None declared.

FUNDING

No sources of funding.

ACKNOWLEDGEMENTS

None.

ETHICAL APPROVAL

No approval is required.

CONSENT

Written informed consent was obtained from the patient's family for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

GUARANTOR

Dr. Michael Bouvet is the guarantor of this paper.

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