

A Challenging Case of Ectopic Adrenocorticotropin Hormone Syndrome with Bronchial Carcinoid and Literature Review

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What is already known on this topic?

Ectopic adrenocorticotropin hormone (ACTH) syndrome is very rare in children. Diagnosis may be delayed because of its rarity. However, early diagnosis is important to prevent comorbidities and improve the patients quality of life.

What this study adds?

Ectopic ACTH cases are mostly reported as individual cases. The case presented here is an example of ectopic ACTH syndrome with bronchial carcinoid. This combination is exceedingly rare in children so all pediatric cases of bronchial carcinoid ectopic ACTH were reviewed. Since this article describes all these cases and the diagnostic process is described in detail, it is hoped this report will be a guide for other colleagues.

Abstract

We report an adolescent boy diagnosed with ectopic adrenocorticotropin hormone syndrome (EAS) due to an atypical bronchial carcinoid. The patient was managed by a multidisciplinary team. He underwent surgery and subsequent chemotherapy and radiotherapy treatments. The patient is still under our follow-up. At the time of writing, eighteen pediatric and adolescent patients with EAS because of bronchial carcinoid tumors have been reported. EAS due to bronchial carcinoids is very rare in children and adolescents. Careful diagnostic evaluation and rapid treatment should be started immediately. Although complete remission is possible, atypical carcinoids have a more aggressive nature. A multidisciplinary approach and follow-up is recommended in terms of quality of life and survival.

Keywords: Ectopic adrenocorticotropin hormone syndrome, carcinoid, Cushing

Introduction

Endogenous Cushing syndrome (CS) is very rare in the pediatric and adolescent periods, resulting from the overproduction of glucocorticoids with an annual incidence rate of 0.7-2.4 cases per million persons (1). CS may be adrenocorticotropin hormone (ACTH) dependent or ACTH independent. Around 80 to 85% of endogenous CS is ACTH-dependent, and of these, 75-80% is caused by ACTH-producing pituitary

adenoma, when it is known as Cushing disease. Ectopic ACTH syndrome (EAS), accounts for almost 15% of all CS cases and occurs when there is production of ACTH from an ectopic source in the body, usually mostly neuroendocrine tumors (NETS) (1,2). ACTH-secreting NETS are usually located in the thymus, lungs, pancreas, or gastrointestinal system, but may also present as Ewing sarcoma, pheochromocytoma, or medullary thyroid carcinoma (3).

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Bronchial carcinoids that arise from bronchial mucosal neuroendocrine cells are the most frequently seen primary malignant lung tumors and also the most common cause of EAS in children (4). These NETS are well differentiated, and the word “carcinoid” distinguishes them from poorly differentiated ones, which include small-cell lung cancer and large-cell neuroendocrine carcinomas. They are subdivided into two groups based on malignancy potential, as typical carcinoids (TC) and atypical carcinoids (AC), and the majority of pediatric patients present with the typical type. The median age of presentation of EAS cases is 9.5 years, with a female predominance (5).

The initial diagnosis does not usually consider CS based on anamnesis and physical examination of pediatric cases. Furthermore, EAS constitutes a very small part of the etiology of CS so the diagnosis and treatment process may be delayed. We discuss a case of an ectopic ACTH-secreting bronchial carcinoid who had been seen in various centers. However, none of these centers had considered CS, despite presenting because of gynecomastia and even having a scheduled gynecomastia procedure planned. In addition, the current literature currently describing pediatric cases of EAS due to bronchial carcinoid is reviewed.

Case Report

A 13.7-year-old boy complained of gynecomastia and excessive weight gain. In his anamnesis, it was learned that he had been examined in different centers for gynecomastia for around one year, and an operation was even planned. There was no prior physical sickness or steroid use. On physical examination, weight was 61 kg [0.55 standard

deviation score (SDS)], height was 157 cm (-0.81 SDS), and body mass index (BMI) was 24.7 kg/m² (0.9 SDS). His blood pressure was 95/65 mmHg (75-90th centile). He had moon facies, truncal adiposity, a buffalo hump, purple striae over the abdomen and breast, hypertrichosis, and gynecomastia (Figure 1). He had pubic hair stage 3 with a stretched penile length of 7 cm and bilateral testicular volume of 10 mL each. The other systemic examination was unremarkable. Biochemical evaluation for liver and renal function tests and lipid profiles were normal. On hormonal evaluation, serum ACTH level was 60.2 pg/mL (NR 7.2-30 pg/mL) and basal cortisol was 20.4 mcg/dL. His midnight and evening serum cortisol concentrations were 19.9 µg/dL and 25.1 µg/dL. CS was confirmed using a low-dose (1 mg) dexamethasone suppression test (LDDST), which revealed non-suppressed blood cortisol (1988 g/dL). Twenty-four-hour urinary free cortisol levels (UFC) were elevated (2760 nmol/24-hour and 3800 nmol/24-hour). A high-dose dexamethasone suppression test (HDDST) also showed no suppression (baseline serum cortisol = 19.2 µg/dL; cortisol after test = 22.1 µg/dL). The serum level of chromogranin-A was elevated (191 ng/mL, NR <100 ng/mL). Dynamic contrast magnetic resonance imaging (MRI) of the pituitary was normal.

Contrast-enhanced computed tomography (CT) of the thorax and abdomen was performed to identify the peripheral ACTH-secreting tumor. This identified a 21x12 mm hypodense homogeneous solid lesion in the left hilus and a 7 mm nodule in the lingula of the left lung with micro mediastinal lymph node enlargements. To further investigate this lesion, ⁶⁸Ga-DOTATATE positron emission tomography (PET)/CT was performed, which revealed pathological activity on the hilus and lingula of the left



Figure 1. Preoperative physical examination findings: moon facies, gynecomastia, purple striae

lung (Figure 2). This finding strengthened the possibility of the lesion being the source of EAS. A biopsy performed on the lesion under endobronchial US revealed findings consistent with a carcinoid tumor (strongly positive ACTH antibody). The patient underwent surgery. The lesion in the hilus was dissected as completely as possible although it was in close proximity to the main bronchus and vascular structures) and removed *en bloc*, and wedge resection was done for the lesion in the left lung. In the histopathological examination of the lung, proliferation consisting of cells with spindle-oval-shaped chromatin in the form of a salt and pepper appearance, showing solid and insular organization,



Figure 2. Positron emission tomography/computed tomography images: lesion in the left lung

was observed. On immunohistochemistry, tumor cells were positive for chromogranin-A, synaptophysin, CD56, thyroid transcription factor-1, and ACTH, suggesting a NETS. The Ki67 proliferation index was 2-3%. Up to 5 mitoses were counted under x10 magnification with pHH3 (phosphohistone H3).

The postoperative first-day ACTH level of the patient decreased to 14.6 pg/mL. The patient was initiated on hydrocortisone replacement (10 mg/m²/day) and monitored every month with morning serum cortisol and ACTH and hydrocortisone was tapered over three months. However, due to incomplete resection of the lesion, radiotherapy and six cycles of chemotherapy (carboplatin and etoposide) treatment were given. In the first year of follow-up, his midnight and evening serum cortisol concentrations were 1.4 µg/dL and 0.55 µg/dL, respectively. A LDDST showed suppressed serum cortisol (0.5 µg/dL). Twenty-four-hour UFC was 41 nmol/24-hours. The pathological activity involvement was reduced in the monitoring ⁶⁸Ga-DOTATATE PET/CT. He had no new metastasis. At his last follow-up (fifteen months postoperatively), he was 15.2 years old, his weight was 47 kg (-1.8 SDS), his height was 171 cm (-0.01 SDS), and his BMI was 16 kg/m² (-2 SDS). He had pubic hair stage 4 with a stretched penile length of 8.5 cm and bilateral testicular volume of 15 mL each. Laboratory examination showed no endocrine abnormalities (growth hormone deficiency, thyroid dysfunction, gonadal suppression, or hyperglycemia). The physical examination findings related to CS, especially gynecomastia, had regressed (Figure 3). Pediatric endocrinology, pediatric oncology, radiation oncology, thoracic surgery, nuclear medicine, and radiology departments continue multidisciplinary follow-up. The endocrinology department performs anthropometric and hormonal (glucose metabolism, ACTH, cortisol,



Figure 3. Postoperative regression in gynecomastia, striae, and cushingoid face appearance

lipid metabolism, bone metabolism, puberty evaluation, and other endocrinopathies that may accompany EAS) evaluations every three months. Oncology and radiation oncology departments use laboratory and imaging methods for monitoring remission or metastasis after chemotherapy and radiotherapy treatments. Nuclear medicine and radiology departments interpret tomography and PET/CT images and jointly decide on the frequency of the tests to be performed.

Literature Search

At the time of writing eighteen pediatric and adolescent patients with EAS because of bronchial carcinoid tumors have been reported in 13 case reports and literature reviews (4,6,7,8,9,10,11,12,13,14,15,16,17). The mean age of the reported patients was 14.1 ± 2.7 years. There were 11 females (61 %) and 6 males (39 %). In one case sex was not disclosed. Two cases were defined as AC. Lymph node metastasis was reported in seven (38.9 %) patients. All of the patients had thoracic surgery, while three had bilateral adrenalectomy operations (Table 1).

There were two more major series of EAS. In the first one, the ages ranged between 8-72 years, and there were 35 patients with bronchial carcinoid tumors. Three deaths and four relapses were reported (18). In the second series, the ages ranged between 12-74 years. There were 81 patients with bronchial carcinoid tumors, and 26 deaths were reported (19). However, the number and outcome of the treatment follow-up of the pediatric cases in these case series were not reported separately from the adult data.

Discussion

CS is very rare in children. The more silent and progressive course of the syndrome in children and the difficulty of the testing often result in a long delay in CS diagnosis. Clinical suspicion based on anamnesis and physical examination is the initial stage in the diagnostic process. Screening and diagnostic procedures for CS evaluate the level of cortisol secretion. These procedures include the late-night salivary/serum cortisol test, the overnight 1-mg LDDST, the 24-hour UFC, and the HDDST (2). The gold standard for identifying hypercortisolemia is to measure cortisol at midnight with an intravenous catheter inserted; a cortisol level exceeding 4.4 mg/dL at that time ensures a high sensitivity and specificity for CS (2). Diurnal testing, however, necessitates an overnight hospital stay and has a limited role in standard screening tests. A serum cortisol $<1.8 \mu\text{g/dL}$ at 0800 h in the morning after LDDST is considered a normal response. The 24-hour UFC threshold value is 90 mcg/24 hours by radioimmunoassay or 50

mcg/24 hours by high performance liquid chromatography/immunochemiluminescence. Anorexia, chronic and severe obesity, pregnancy, chronic activity, depression, poor diabetes control, anxiety, malnourishment, and too much water consumption are all pseudo-Cushing states that may result in false-positive elevations during 24-hour UFC measurements. All of these tests have limits, and additional tests are typically required to confirm the diagnosis because none of them has 100 % diagnostic accuracy (2). In the presented case, firstly, the diagnosis of CS was confirmed by demonstrating elevated midnight cortisol, indicating a lack of diurnal rhythm, poor suppression in the LDDST, and elevated 24-h UFC. Once the CS diagnosis was confirmed, serum ACTH level should be evaluated to distinguish ACTH-dependent (Cushing disease or ectopic ACTH) and ACTH-independent CS. Children with an ACTH-dependent type of the syndrome can be identified with a sensitivity of 70 % using a spot morning plasma ACTH level of at least 29 pg/mL (2). The patient had a high serum ACTH level and thus had ACTH-dependent CS.

EAS in children is much less common than in adults (4). Diagnosis may be challenging. In both children and adults, the diagnostic procedure to distinguish EAS from a pituitary adenoma is the same (2,5). Cushing disease and EAS can be distinguished using the desmopressin or corticotropin-releasing hormone stimulation test, the HDDST, and bilateral inferior petrosal sinus sampling (BIPSS). Combining the blood tests with MRI is a non-invasive strategy (2,5). Hypophyseal and cerebral MRIs were performed because pituitary adenoma in children is the main cause of ACTH production. In ACTH-dependent CS, whole-body CT should be performed after hypopituitary imaging to seek an ectopic cause (2,5). In the present case, CT of the thorax and abdomen showed a solid lesion in the left lung after normal hypopituitary MRI results. It may be challenging to pinpoint the location of the ACTH-secreting tumor, and a single positive imaging study may be a falsely positive result. An octreotide scan may be used to validate the diagnosis of EAS (2,4). According to a recent meta-analysis, both ^{68}Ga -DOTA-peptide and ^{18}F -fluorodeoxyglucose (FDG) are very sensitive for identifying pulmonary carcinoids, but ^{68}Ga -DOTA-peptide is more sensitive than ^{18}F -FDG (90.0 % vs. 71.0 %) (20). In our case, ^{68}Ga -DOTATATE PET/CT revealed pathological activity at the hilus and lingula of the left lung. Despite having the highest sensitivity and specificity, BIPSS was not necessary for our patient since we were able to make the correct diagnosis using precise, concordant biochemical and radiological investigations.

Biopsy confirmed a carcinoid tumor, and the pathologic examination showed it to be an AC tumor. The most frequent primary malignant lung tumor in children is

Table 1. Review of literature on bronchial carcinoid tumors causing ectopic adrenocorticotropin hormone syndrome

Reference	Year	Age	Sex	Type	LN metastasis	Treatment	RT	Outcome	Recurrence
Ward et al. (7)	1983	15	F	BC		Metyrapone + surgery		6 th month control normal	
Magiokou et al. (8)	1994	11	F	BC		Bilateral adrenalectomy + surgical resection			
Weber et al. (9)	1995	17	F	BC	+	Metyrapone + surgery	+	Well after 5 years follow-up	
Ilias et al. (18)	2005	8-72 years	35 patients	BC	+ in 18 patients	Preoperative treatment? + surgery + CT in 6 patients	6 patients	Death in 3 patients	In 4 patients
More et al. (10)	1988	18	M	Atypical BC	-	Ketoconazole + mitotane + bilateral adrenalectomy + surgery		Well after 16 years follow-up	-
	1995	15	M	Typical BC	+	Surgery		Well after 4 years follow-up	
	1995	17	F	Typical BC	+	Ketoconazole + mitotane + surgery		Well after 16 years follow-up	4 times
	2005	16	M	Typical BC	+	Ketoconazole + surgery		Well after 3 years follow-up	-
Dias et al. (11)	2006	NR	NR	BC					
Bhansali et al. (12)	2009	NR	M	BC		Surgery + recurrence: bilateral adrenalectomy + CT	+	Alive	1 times
		10	M	BC		Surgery - ARDS		Death	
Kakade et al. (13)	2013	9	F	BC					
		14	F	BC	-	Surgery		Alive	
Goldberg et al. (14)	2014	15	F	Atypical BC	+	Surgery		Well after 1 year follow-up	-
Karageorgiadis et al. (15)	2015	15,5	F	BC	+	Surgery (2 times)			1 times
Banerjee et al. (16)	2015	14	F	BC	+	RT + CT + temezolomide + mifepristone		Death	
Potter et al. (5)	2019	13	F	BC		Surgery			
Saxena et al. (4)	2019	12	F	Typical BC	-	Surgery		Well after 1 year follow-up	
Golounina et al. (19)	2021	12-74 years	81 patients	BC		Surgery		Death in 26 patients	
Attri et al. (17)	2023	17	M	BC		Surgery		Structural remission	

LN: lymph node, RT: radiotherapy, F: female, M: male, BC: bronchial carcinoid, CT: chemotherapy, NR: not reported, ARDS: acute respiratory distress syndrome

bronchial carcinoid, and 4 % of these tumors are associated with CS. Histopathological analysis supports the diagnosis. Depending on the presence or absence of necrosis and an increased mitotic index (>2 mitoses/HPF), they are categorized as atypical (19%) and typical (90%) (4,6). Biomarkers such as synaptophysin and chromogranin A may be positive in either kind. All bronchial carcinoids are best treated surgically, and when feasible, lung-sparing resections (such as wedge, segment, or sleeve resections) are advised for children and adolescents (6,21). Lymph node resection is recommended in both types, but is important especially in AC because of their malignant potential (4). Somatostatin-based treatment, cytotoxic chemotherapy, and/or radiation should all be considered in cases with unresectable tumors (4,22). In the present case, complete resection was not possible, but lymph node resection was done, as recommended in the literature. Inhibitors of steroidogenesis, such as metyrapone and ketoconazole, as well as anti-glucocorticoid medications (mifepristone), can be used to treat hypercortisolemia when contraindication of surgery is present or when the patient has not recovered from surgical resection after surgery (4,23). The tumor's size, lymph node status, and histology all affect the prognosis. AC tumors have a worse 5-year survival rate of 60-75 % in pediatric series, while this is 88-92 % for TC tumors (4,6,21). In a study by Degnan et al. (22), aggressive characteristics of ACs were shown in the pediatric cohort, and two of the five bronchial carcinoids were shown to have a higher prevalence of metastatic illness. Bronchial carcinoid tumor recurrence was reported in 10 % of cases in an investigation of the National Cancer Database ($n = 3335$) (3 % in TC and 25 % in AC) (24). Lou et al. (25) reported that post-resection recurrence rates were 5 % for TC and 20 % for AC.

The hypothalamic-pituitary-adrenal axis may be inhibited for up to a year following surgery for Cushing disease. After removal of tumors causing CS, including EAS, glucocorticoid replacement treatment is thus for up to a year, and occasionally even longer (23,26). The duration of tertiary adrenal insufficiency may vary depending on the origin of the condition: it was shortest in cases of ectopic CS, intermediate in cases of Cushing disease, and longest in cases of adrenal CS brought on by cortisol-producing adenoma (27). In the present case, we were able to taper and later cut the hydrocortisone treatment within 3 months.

Patients being treated for AC or TC with positive lymph node involvement should have CT surveillance (24). The sensitivity of ^{68}Ga -DOTATATE for ectopic CS location in diagnosis is high for both occult primary tumors and metastatic lesions (28). However, there is an ongoing debate over the use of PET/

CT for assessing tumor response to therapy. This is because lower uptake on PET/CT may suggest a decrease in tumor volume, but this is only true for well-differentiated NETs that are positive for the somatostatin receptor (SSR). Poorly differentiated SSR-poor tumors are challenging to visualize on ^{68}Ga -DOTATATE PET/CT, but are typically well seen on FDG PET/CT due to their strong glycolytic metabolism (29).

Although a change in tumor size is a good indicator of true response, no decrease in size does not necessarily indicate that there has been no response to treatment. Some lesions may enlarge as a result of cystic or liquefied necrosis that develops following successful therapy. If imaging is carried out a few weeks or months following therapy, such structural alterations are more frequent and may deceive the decision-maker. Radiologists should also be aware that increased cellular expression of SSR in a variety of physiological and other pathologic processes, such as the activity of the pancreatic unsinate process, epiphyseal growth plates, reactive nodes, degenerative bone disease, and changes following radiation therapy, can cause interpretation errors (29,30). Combined with anatomical imaging (CT or MRI), it is the gold standard functional imaging modality for evaluating well-differentiated NETs (28,29).

Conclusion

EAS caused by bronchial carcinoids is very rare in children and adolescents. Careful diagnostic evaluation is important and treatment should be started immediately. Although complete remission is possible in bronchial carcinoids, ACs tend to have a more aggressive nature. A multidisciplinary approach and follow-up will improve quality of life and survival.

Ethics

Informed Consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal.

Footnotes

Authorship Contributions

Surgical and Medical Practices: Sema Nilay Abseyi, Zeynep Şıklar, Elif Özsu, Zehra Aycan, Nurdan Taçyıldız, Ayten Kayı Cangır, Merih Berberoğlu, Concept: Sema Nilay Abseyi, Zeynep Şıklar, Merih Berberoğlu, Design: Sema Nilay Abseyi, Zeynep Şıklar, Merih Berberoğlu, Data Collection or Processing: Sema Nilay Abseyi, Analysis or Interpretation: Sema Nilay Abseyi, Literature Search: Sema

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