World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(CASE REPORT)

Corticoadrenaloma: A case report from the pediatric haemato-oncology and pediatric endocrinology departments at the Hassan II University Hospital of Fez

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World Journal of Advanced Research and Reviews, 2024, 22(01), 1882–1886

Publication history: Received on 20 March 2024; revised on 28 April 2024; accepted on 30 April 2024

Article DOI: https://doi.org/10.30574/wjarr.2024.22.1.1336

Abstract

Adrenocortical carcinoma is a rare tumor, representing 0.2% of pediatric cancers. Diagnosis is made on a combination of clinical, biological, radiological, and anatomopathological arguments. Treatment involves tumor excision surgery, sometimes supplemented by chemotherapy for forms with a high risk of recurrence. Prognosis is often reserved, with a non-negligible death rate.

This report presents three cases of adrenocortical carcinoma treated at our institution between 2017 and 2023. The patients were three girls aged 5 years, 8 months, and 11 years, respectively. They were admitted for signs of hyperandrogenism and arterial hypertension. The tumor was left adrenal in all patients. The histological score was Severe Weiss 6 or 7. All patients underwent tumor resection, and they all benefited from this procedure. Despite chemotherapy in one patient, normalization of blood pressure figures postoperatively, two patients progressed to recurrence and subsequently died. The objective of this study is, through three observations, to describe the problem of the management of adrenocortical carcinoma (ACC) on the one hand, and to review the literature on the management and the prognosis of corticosteroid ACC in children.

Keywords: Corticoadrenaloma; Adrenal; Cortisol; Virilization; Cushing

1. Introduction

1.1. The adrenal gland is located above each kidney and is composed of two distinct areas

The outer layer is the adrenocortical zone, which secretes different hormones. The glucocorticosteroids, which include cortisol, act on blood sugar. Mineralocorticoids, represented by aldosterone, regulate sodium and potassium balance in the body. Androgens also play a role. The inner layer is the adrenal medulla, which secretes adrenaline and noradrenaline. These hormones affect the rhythm of the heart, the contraction or dilation of blood vessels, and blood pressure.

A variety of tumors may develop in this region, including benign (more frequent) and malignant ones. Among these, pheochromocytoma, which develops in the spinal cord area, occurs in nine cases out of ten, while corticosteroidaloma develops at the cortical zone.

Adrenocortical carcinoma is particularly rare in children. Its incidence is estimated at 1.3% in the USA, or 0.2% of pediatric cancers (1). It is sometimes found in the context of a predisposition syndrome, such as Beckwith-Wiedemann

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or Li and Fraumeni syndromes. The diagnosis is often made on a combination of clinical, biological, radiological, and anatomopathological arguments. Their treatment involves tumor excision surgery, sometimes supplemented by chemotherapy, which is most often based on cisplatin, etoposide, and doxorubicin, combined with mitotane. For forms with a high risk of recurrence, this treatment is often recommended. The role of radiotherapy is a topic of particular interest, particularly in light of the frequent involvement of abnormalities of TP53. The prognosis is often unfavorable, with a notable mortality rate.

The objective of this study is, through three observations, to describe the problem of the management of adrenocorticaloma on the one hand, and to review the literature on the management and the prognosis of corticosteroidoma in children.

1.2. Observations

1.2.1. Observation 1

DK aged 5 years, no particular ATCD, present for 2 months, weight gain, acne, hursitism, hypertension (18/11 cm Hg) with signs of virilization (cushingoid morphology). The hormonal balance was elevated, with a cortisol level of 24.4 µg/dL, DHEAS 285 ng/mL, testosterone 1.10 ng/mL, ACTH 3.7 pg/mL, potassium 2.8 mmol/L, and blood sugar 3 g/dL.

Abdominal CT revealed a large mass measuring 72*65*65 mm at the level of the left adrenal gland, with no sign of distant invasion.

The patient underwent tumor resection without intraoperative incident and simple postoperative follow-up. The pathological examination concluded with an adrenocortical syndrome, with a Weiss score of 7. The patient died after the first session of chemotherapy in a state of multivascular failure.

1.2.2. Observation 2

HB, aged 8 months, from a first-degree consanguineous marriage, and death of his older brother at the age of 4 from an undetermined cause. The patient presents with obesity, high blood pressure (potentially reaching 18/12 mmHg), pubic hair, hirsutism, hoarseness of the voice, signs of virilization, and a Cushingoid morphology.

The hormonal balance is disturbed, with serum cortisol at 29 ug/dl, DHEAS at 1432 ng/ml, testosterone at 0.5 ng/ml, ACTH at 3.9 pg/ml, and potassium at 3.

Abdominal CT revealed a large mass measuring 45*38*44mm in the left adrenal region, with no sign of distant invasion. The patient underwent tumor resection without intraoperative incident and simple postoperative follow-up with normalization of blood pressure figures. The pathological examination concluded with an adrenocortical syndrome with a Weiss score of 6. The family was referred to the oncology department for further treatment, but they never showed up. The patient was admitted two years later as an emergency due to febrile respiratory distress and hypertension, hyperglycemia. Thoracoabdominopelvic CT revealed a local recurrence with pulmonary metastases. The patient subsequently died.

1.2.3. Observation 3

KD is a 11-year-old individual with an undocumented history of convulsive seizures. The patient was admitted for obesity, acne, large stretch marks on the thighs and buttocks, hair growth in the pubic area with clitoral hypertrophy, and a BP reading of 20/13 mm Hg, which is considered to be a threat to the patient's health. The hormonal balance was disrupted, with serum cortisol at 13.4 µg/dL, testosterone at 25.50 ng/mL, ACTH at 2.9 pg/mL, and potassium at 4. Abdominal CT revealed a large mass measuring 46*32*47 mm at the level of the left adrenal gland, with no evidence of distant invasion. Following stabilization of the patient's blood pressure, tumor resection was performed without incident. The patient's postoperative course was uncomplicated, with normalization of blood pressure figures. The atomo-pathological examination revealed that the tumor was an adrenal cortex, with a Weiss score of 7. Two years later, the patient was readmitted due to the recurrence of hypertension. A thoracoabdominal pelvic CT scan showed a local recurrence with hepatic and pulmonary metastases. The patient benefited from surgery and subsequently died 48 hours after the procedure.

2. Discussion

The objective of this study is, through three observations, to describe the problem of the management of adrenocorticaloma on the one hand, and to review the literature on the management and the prognosis of corticosteroid enaloma in children.

This study confirms the rarity of adrenocortical carcinoma, with only three cases over seven years, representing a frequency of 0.05% of all hospitalizations in the pediatric hemato-oncology department. Over a period of 11 years, 13 cases of corticoadrenaloma were recorded by Nonato M. et al. (2). Adrenocorticaloma ranks third among adrenal tumors, behind neuroblastoma and gangioneuroma (3).

It is primarily observed in females during the first decade of life (1,4,5,6,7,8,9). Adrenocortical tumors may be either secreting or non-secreting. Secreting tumors represent approximately 60% (10). In this case, the manifestations of hypercortisolism are prominent.

When the tumor is non-secreting, local signs such as abdominal pain are prominent (11). The symptoms reported by our patient were consistent with those of secreting adrenocortical tumors. This is characterized by a clinical polymorphism, including Cushing's syndrome, hypertension, signs of virilization in girls or feminization in boys, and so on. This triad was present in all our patients. However, this clinical picture may be incomplete due to the presence of a single sign in certain cases (12). The basic hormonal assessment includes cortisol, DHEAS, testosterone, ACTH, and then biochemistry including potassium and blood sugar. This assessment is disturbed in a variable way. An increase in DHEAS is reported in 84%, testosterone in 89%, and cortisol in 46.7% (13, 14).

With regard to imaging, abdominal CT is useful in determining the orientation of an abdominal mass, specifying its characteristics, its location, and its loco-regional relationships (organic and vascular). It also allows for the identification of the mechanisms of tumor spread. These elements are of particular importance in assessing the operability of the tumor and guiding the surgeon during tumor resection, which is the cornerstone of the treatment of adrenocortical tumors.

In light of the clinical context of hypertension, the presence of clinical and biological hyperandrogenism, and the absence of other potential diagnoses, the most likely diagnoses to be considered are adrenocortical carcinoma, pheochromocytoma, and adrenal neuroblastoma. The anatomopathological examination allows for the diagnosis to be corrected.

None of our patients exhibited metastases at the time of diagnosis or signs of locoregional invasion. However, 15% of metastases at the time of diagnosis is reported in the literature, with the majority occurring in the lungs and liver (13,15,16).

Surgical excision of the tumor is the primary treatment modality. The procedure is typically performed via laparotomy, through a transperitoneal route after an ipsilateral subcostal incision. In adults, we employ laparoscopy as an alternative approach. However, this method is not recommended in children due to the friable nature of Cotricosurrenalomas. The incidence of intraoperative tumor rupture has been evaluated in pediatric series at 20% at the time of initial excision, and greater than 40% in the event of surgical intervention on a local recurrence (9,13,14).

Chemotherapy and radiotherapy are recommended for patients with an unresectable tumor, at high risk or with positive resection margins (17).

Moreover, adjuvant chemotherapy has not demonstrated efficacy in improving patient survival (10,18). Zancanella et al. (13) reported a small prospective series of 11 children treated with the combination of cisplatin, etoposide, doxorubicin in combination with mitotane, noting two complete responses and five complete responses. Minor or partial responses were also observed. This therapeutic combination is currently the most commonly used in children. The efficacy of this treatment is currently being evaluated in an international prospective study conducted by the team at St. Jude Children's Research Hospital for the Children's Oncology Group (protocol ARAR0332). In this protocol, the treatment is planned to be administered to patients with stage III or IV adrenocortical carcinoma (9.13).

2.1. Prognosis

The prognosis for an adrenocortical tumor depends on whether the tumor was completely resected during surgery. The five-year overall survival rate for adrenocortical carcinoma is 85% if the tumor is completely removed or totally

removed by surgery. However, if the tumor is not completely resected, the five-year overall survival rate is less than 40%. Factors that influence the chances of recovery include:

The size of the tumor, the extent of the initial surgery, the age of the patient, and the histological characteristics of the tumor all influence the prognosis of adrenocortical carcinoma. Despite treatment efforts, the prognosis of adrenocortical carcinoma is poor. In the series of eight cases of adrenocortical carcinoma by J. Li et al. (11), more than 50% of patients were diagnosed with metastatic disease, including pulmonary, hepatic, and lymph node metastasis. Tumor recurrence has been reported even in cases of guard II after complete tumor section and chemotherapy at approximately one year (11). For a Weiss score > 6, the estimated average survival is 6 months (11).

All patients exhibited a Weiss score of at least 6, which may have contributed to the unfavorable prognosis observed in all cases.

3. Conclusion

Adrenocortical tumor is a rare malignant tumor. When it is secreted, the manifestations of hypersecretion of corticosteroids help to guide the diagnosis more quickly. The treatment is primarily surgical with a wide cardiological resection. Despite treatment efforts, the prognosis is poor with recurrence and death, which makes it difficult to obtain prospective data, particularly with regard to therapeutic management. Most of the data are inhomogeneous, coming from small retrospective series. The rarity of the disease and the poor prognosis of locally advanced and metastatic forms justify the systematic registration of these patients and the harmonization of therapeutic care at the national level or even internationally.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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