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Corticosurrenaloma: About a Pediatric Case

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Abstract

Introduction: This study aims to describe the outcome of adrenocortical cancer in children through observation. **Observation:** A 10-year-old girl with no previous pathological history. She presented headaches and severe hypertension with clinical and biological signs of hyperandrogenism for 6 months. An abdominal CT scan showed an encapsulated left adrenal mass without local or regional invasion or secondary location. A pheochromocytoma or adrenal neuroblastoma was first suspected. The blood pressure was stable at 130/65 mmHg under antihypertensive drugs. She underwent a complete tumor resection without any intraoperative incident. The pathologic study confirmed the adrenocortical carcinoma scored Weiss 7. The severe high blood pressure reappeared 2-year later despite antihypertensive drugs. The thoracic abdominal and pelvic CT scan showed a locally advanced tumor recurrence in the left adrenal gland with parenchymal nodes in the lungs and liver. The outcome was fatal despite medical and surgical management. **Conclusion:** Adrenocortical cancer is a rare tumor. It is important to hormonal testing in the presence of Cushing's syndrome in children. It can give a strong indication of the diagnostic possibilities. Histology confirms the diagnosis. The evolution is covered by complications, in particular recurrence with life-threatening metastases.

Keywords

Adrenal Tumor, Adrenal Cortex, Hyperandrogenism, Child

1. Introduction

Adrenocortical carcinoma represents 0.2% of malignant tumors in children with an incidence of 0.2 to 0.3/1,000,000 people [1]. Clinical manifestations vary depending on whether the tumor is secretory or not. Surgical treatment is current-

ly the only radical treatment [2]. Despite management efforts, the prognosis remains poor.

2. Case Report

A 10-years old girl with no previous pathological history. She presented since 6-months moderate headache, axillary hyper-sweating, unexplained facial-trunk weight gain (65 kg), with hyper hirsutism and stretch marks. Admitted in paediatrics for malignant high blood pressure varying between 160/100 and 180/120mmHg. Biological findings included 17-B estradiol 25 nmol/l (very high), FSH < 0.05 IU/L (very low), HLH 0.02 (normal), cortisol 13.4 microgram/dl (normal) urinary cortisol 199 microgram/dl (normal < 30), testosterone 25.25 ng/ml (normal < 0.3), dehydroepiandrosterone sulphate 0.4 ng/ml and normal urinary catecholamines. The rest of the preoperative work-up was normal.

An abdominal CT scan revealed a large left adrenal mass measuring more than 10 cm and encapsulated, with no signs of local or regional invasion or secondary location. Pheochromocytoma should be considered first without completely ruling out adrenal neuroblastoma.

A cerebral MRI was performed: in search of a central origin, which was anomal.

She had blood pressure monitoring, and put on a calcium channel blocker (amlodipine) and a beta-blocker (control). Good clinical evolution with the blood pressure stable at 130/65mmHg.

She underwent uneventful complete surgical resection. Discharge after 7-days under antihypertensive treatment with instructions to do local care every day.

The histological study confirmed a Weiss score of 7 for adrenal carcinoma. Radiological-histological conclusion: Our patient presented with ENSAT stage II adrenocortical carcinoma and a Weiss score of 7 [3].

The patient had a good clinical course with stabilisation of blood pressure.

Blood pressure returned to 170/120 two years later.

An abdominal and pelvic CT scan revealed a local and advanced tumor recurrence in the left adrenal gland with pulmonary parenchymal nodules. After multidisciplinary consultation, she underwent uneventful surgical resection. The mid-term evolution was marked by death in a multi-visceral failure.

3. Discussion

Corticosurenaloma is a rare malignancy tumor. During 11 years, 13 cases of adrenal corticosurenaloma have been recorded by Nonato M. *et al.* [1]. This case is the first observation of our service with almost 20 years of experience.

Adrenocortical cancer is the third most common adrenal tumor after neuroblastoma and ganglioneuroma [4].

The onset of symptoms varies between 1.6 and 12 years of age, with a female predominance [1] [5]. Adrenocortical cancer can be secretor or non-secretory. Secretor form accounts for 60% [6]. In this case, the manifestations of hypersecretion of corticosteroids are in the foreground. In the non-secretor form, local

signs such as abdominal pain are in the foreground [5].

The symptoms reported by our patient were those reported in over secretion of the adrenal cortex.

High blood pressure is reported at 38.5% [1]. This one, associated with sweating, is highly confusing with pheochromocytoma. High blood pressure, paroxysmal sweating, headache, and palpitation are the main symptoms of pheochromocytoma.

As in our patient, the corticosurrenaloma is located on the left in more than half of the cases [1].

Pituitary MRI was performed in our study to rule out a central origin of hyperandrogenism. It was normal.

Given the clinical context of hypertension and signs of clinical and biological hyperandrogenism; the diagnoses evoked were: adrenal carcinoma, pheochromocytoma, and adrenal neuroblastoma.

Anatomopathology examination confirmed the diagnosis of corticosurrenaloma.

Urinary catecholamines were negative. It makes the pheochromocytoma and adrenal neuroblastoma less doubtful, without formally excluding them.

Currently, tumor resection is the only radical treatment [2] [6]. Located on the right side, an enlarged nephrectomy can be performed.

On the left side, a caudal splenic-pancreatectomy associated with or no to nephrectomy can be performed [6].

Chemotherapy and radiotherapy are recommended for patients with unresectable tumours, high risk, or positive resection margins [2].

In addition, adjuvant chemotherapy has not been proven to improve patient survival [6] [7]. In our case, the patient did not initially present with metastases, which motivated the choice not to undertake chemotherapy. In the immediate after the blood pressure remained stable.

Despite management efforts, the prognosis of adrenocortical cancer is poor. In the series of eight cases of adrenocortical cancer by Li J. *et al.* [5], more than 50% of the patients were at the time of diagnosis at the stage of metastasis, including lung, liver, and lymph node. For a Weiss score > 6, the average survival is estimated at 6 months [5]. Tumour recurrence is reported even for guards II after complete tumor resection and chemotherapy at about one year [5].

Our patient had symptoms suggestive of localized adrenocortical cancer confirmed by a histological study. Primary resection resulted in improvement of symptoms, but after two years of stability, they recurred. The histological score of Weiss 7 which implies venous invasion could explain the local recurrence with simultaneous distant metastases.

The fatal outcome was secondary to advanced tumor progression with lung and liver metastases,

4. Conclusion

Adrenocortical cancer is a rare malignancy. When it is secretory, the manifesta-

tions of hypersecretion of corticosteroids help to orientate the diagnosis more rapidly. Treatment is primarily surgical with extensive cardiac resection. Despite management efforts, the prognosis is poor with recurrence and death.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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