

Malignant Adrenal Cortex in Children: About 6 Cases

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Abstract

Original Research Article

The pediatric adrenocortical carcinoma is a rare cancer estimated at 0.3-0.38 cases per million children per year. Its diagnosis is based on a set of clinical, biological and radiological arguments. Its rarity, coupled with a relatively poor literature, especially from a prospective point of view, make the therapeutic approach to this cancer a real challenge. This work identifies 6 patients who were treated in the pediatric surgery department A of the children's hospital in Rabat over a period of 28 years, and aims to shed light on the various clinical, biological and radiological aspects of malignant adrenocortical carcinoma, as well as different therapies used and the outcome of these children; and offers a comparative approach with what the literature offers.

Keywords: Adrenocortical carcinoma, children, virilization, surgery.

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INTRODUCTION

Malignant Adrenal cortex is a rare endocrine tumor of the adrenal cortex. Its incidence is approximately one to two new cases per million per year. With a 5-year survival rate of 20% at 35% [1]. Most frequently, the diagnosis is made when there are signs of hormonal hypersecretion whose symptomatology is variable, depending on the route of hormonal synthesis developed by tumor endocrine tissue and / or before a tumor syndrome. Her early stage tumor resection is an element of good prognosis.

Through this work reporting 6 cases of malignant adrenal cortex in children caught in charge between 1992 and 2020, in the pediatric surgery department A at the children's hospital in Rabat, high lighting the different presentations clinical, biological and radiological findings, as well as the various in charge proposed.

MATERIAL AND METHODS

This is a retrospective descriptive study of six cases presenting a malignant adrenal cortex, collected from the surgical department A in the hospital of children in Rabat, spanning from January 1992 to July 2020. Data collection was carried out from patient records.

All our patients were first explored by a hormonal assessment (testosteronemia, DHEA, ACTH, 8h cortisolemia) the abdominal scanner was performed in all our patients in addition to the abdominal ultrasound.

The metastatic assessment was performed in three patients including a chest CT scan and a bone scan.

RESULTATS

The age of the patients ranged from 11 months' predominance. A strong female predominance has been reported, with 5 girls for 1 boy. 5 cases had a right localization.

All the patients presented a secreting form, and all exhibited signs of virilization; only 2 showed signs of Cushing's syndrome, and 2 presented a mixed form. No case of Conn syndrome, feminizing or non-secreting form has been found, three patients presented with metastases: they were exclusively hepatic and/or pulmonary.

All patients received a CT scan as part of the diagnosis, 5 of whom also received an ultrasound. 5 patients received surgery, 4 of whom received adjuvant chemotherapy. No patient received mitotane. The 6th

patient was lost to follow-up before treatment. The Weiss scores found in these patients were all ≥ 3 , including three ≥ 5 . Of the 5 operated patients, 2 died within a year of surgery, 2 were considered to be in complete remission, and the follow-up of the last patient is still ongoing.

DISCUSSION

Malignant Adrenal is a malignant tumor that develops in depends on the adrenal cortex. It represents less 1% of pediatric cancers. It's a pathology that affects different age groups, it is more common in children under 5 years old, and in adults around the 4th - 5th decades, with predominantly female [2]. Which agrees with our study including 5 cases under 5 years old with a female predominance.

Clinically, the symptomatology may result from a secreting, non-secreting or metastatic form. Secreting adrenal cortex is the most common form in children with an estimated frequency of 95% [2]. In our study, our 6 patients presented with a secreting form. The clinical picture varies depending on the type of secretion. One functional adrenal cortex can thus be revealed by virilization from a distance the most frequent form, or by a cushing syndrome; more rarely by a Conn syndrome or a feminization table, without forgetting the forms mixed [3].

In our series, the 6 patients had pubic hair developed, all 5 daughters presented with hirsutism and hypertrophy clitoral, and the only boy had a developed penis. The acne was present in 3 patients, hoarse voice in 2 patients, and a greater than 2 SD in 2 patients.

Cushing's syndrome was present in 2 patients, no cases of Conn's syndrome were noted mixed forms are characterized by the presence of at least two types of secretions hormonal. It should be noted that the association of Cushing's syndrome and virilization in children is often a sign of malignancy [4, 5]. In our series, 2 patients presented with a mixed form.

The non-secreting form is characterized by hormone production at a rate that does not lead to clinical repercussions [6], thus causing a delay in diagnosis [7], the metastatic form is relatively common at the time of diagnosis. Through the various studies, metastases seem to be localized mainly at hepatic and pulmonary levels, and more rarely at the level other locations: bone, pancreatic, uterine, cutaneous, and cerebral.

This agrees with our study, where the 3 patients who presented metastases at the time of diagnosis or during follow-up had localizations hepatic and / or pulmonary.

The hormonal assessment of patients with suspected tumors of the adrenal cortex explains the clinical symptoms and should include the following blood strengths: cortisol, testosterone, dehydroepiandrosterone sulfate (DHEA-S), androstenedione, 17-hydroxyprogesterone, aldosterone, plasma renin activity, deoxycorticosterone and corticosterone [8], the literature confirms the rarity of the exclusivity of the aldosterone production [9].

Abdominal CT scan and abdominal magnetic resonance imaging are the two reference exams [3, 10, 11, 12]. The CT scan reveals a large, heterogeneous tumor due to the presence of intratumoral areas of bleeding, necrosis or calcifications (Fig 1). Injected CT is also an excellent test to assess loco-regional and remote extension, allowing precise staging, highlighting para-aortic and hepatic lymph node metastases (Fig 2) pulmonary, and bone. MRI allows an excellent study of loco-regional relations hips (Fig 3), and also of vascular invasion, in particular of the inferior vena cava.



Fig 1: CT appearance of a right malignant Corticosurrenaloma

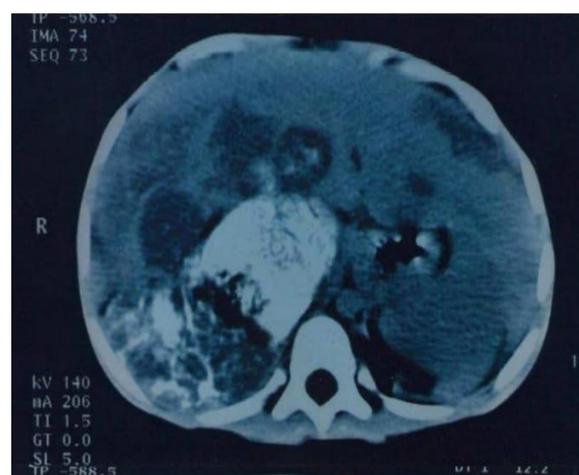


Fig 2: CT appearance of a right adrenocortical carcinoma with hepatic metastases

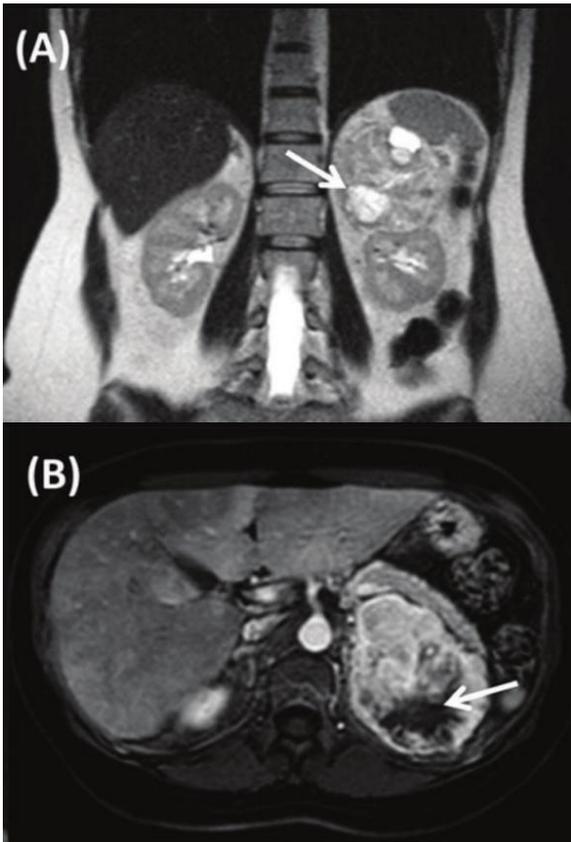


Fig 3: MRI appearance of a left adrenal cortex

On the histological level (Fig 4 & 5) The diagnostic difficulty occurs when the tumor is intrasurrenal, which is the case in pediatric forms [13, 14]. This is how many multiparametric systems have been developed and widely used is that proposed by Weiss *et al.*,

This is based on nine microscopic criteria and the sum of the criteria present defines a score ranging from 0 to 9. The presence of at least three criteria is in favor malignancy [15].

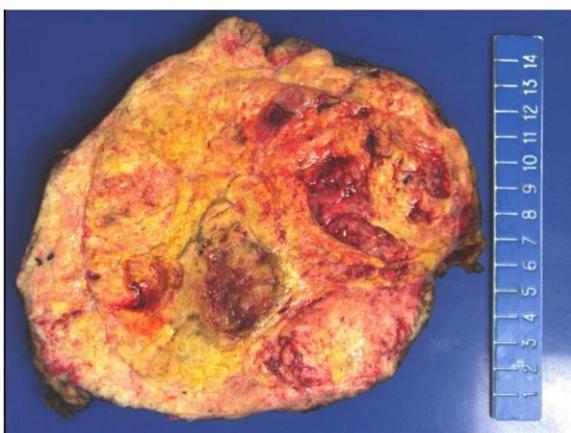


Fig 4: Macroscopic section of a malignant adrenocortical carcinoma

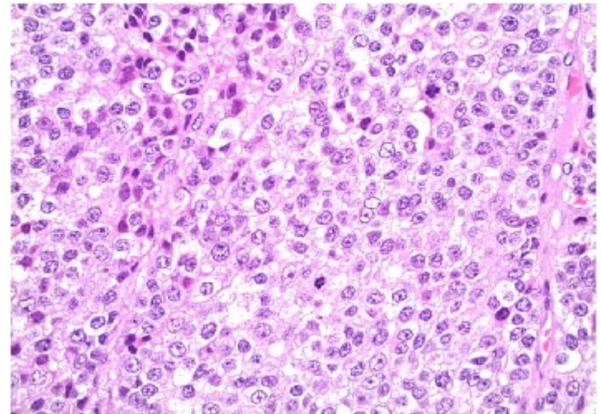


Fig 5: Histological section of a malignant adrenal cortex showing numerous interrupted mitoses

The first-line treatment for malignant adrenocorticals, regardless of the type of secretion is surgery resulting in remission in 0 to 50% of cases depending on the stage during the intervention [16]. The most recommended surgical approach, especially in children, the anterior laparotomy remains. The incidence of intraoperative tumor ruptures has been assessed in pediatric series at 20% at the time of excision initial, and greater than 40% in the event of an intervention surgery on loco regional recurrence [13, 17].

The tumor must therefore be handled with great precaution during surgery. The occurrence of a rupture tumor is associated with a poor prognosis, which advocates for offering systematic adjuvant treatment to these patients [13]. Given the impact of surgery on survival, it is necessary to assess the possibilities of excision surgical metastases, especially pulmonary as needed after neoadjuvant chemotherapy as well only in the event of local and / or metastatic recurrence. In indeed, it has been shown that these exereses sometimes multiple, if they were macroscopically complete, significantly improved survival overall patients [18, 19].

The type of gesture indicated and possible resection are determined beforehand based on the staging of adrenal cortex, mitotane, chemotherapy, and radiation therapy, not to mention targeted therapies which are currently mainly in the field of research, have a variable place according to the authors.

CONCLUSION

Malignant adrenocorticals are tumors rare in children and sometimes fit into the framework predisposition syndromes such as syndromes by Li and Fraumeni or by Beckwith-Wiedemann. Their diagnosis is often based on a bundle of arguments clinical, biological, radiological and anatomo-pathological. Their treatment involves surgery tumor resection, sometimes supplemented by chemotherapy

The role of radiotherapy is discussed. The rarity of the disease and the poor prognosis locally advanced and metastatic forms justify the systematic registration of these patients and harmonization of therapeutic care.

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