International Journal Dental and Medical Sciences Research
Volume 7, Issue 4, July – Aug. 2025 pp 14-17 www.ijdmsrjournal.com ISSN: 2582-6018

Pediatric Adrenal Masses beyond Neuroblastoma: A Retrospective Study of 7 Cases and Literature Review

Mouna Lazrak(1),HidayaZitan,MeryemHadir, MounimOchen , Jawad Bouljrouf, Mounir Kisra

Department of Pediatric Surgery, Children's Hospital of Rabat – Ibn Sina University Hospital, Morocco

Date of Submission: 01-07-2025 Date of Acceptance: 10-07-2025

ABSTRACT

Objective: To analyze the clinical, diagnostic, and therapeutic characteristics of non-neuroblastoma adrenal masses in children through a series of 7 cases managed at a tertiary center.

Methods: A retrospective descriptive study of 7 children followed between 2016 and 2021 at the pediatric surgery department of the Rabat University Hospital for adrenal masses excluding neuroblastoma.

Results: Five cases were adrenocortical carcinomas (including four with androgenic secretion) and two were pheochromocytomas. The average age was 6 years (ranging from 7 months to 14 years). The diagnosis was guided by imaging (CT scan, MRI) and confirmed by histopathological examination. All children underwent surgical resection, and adjuvant chemotherapy was administered for malignant adrenocortical carcinoma cases. The outcome was favorable in most cases (one suspected local recurrence).

Conclusion: Pediatric adrenal masses unrelated to neuroblastoma are rare. Their management relies on complete surgical excision, ideally in a specialized setting, supported by histological analysis and a multidisciplinary approach.

Keywords: Adrenocortical carcinoma, pheochromocytoma, child, adrenal mass, pediatric surgery.

I. INTRODUCTION

Adrenal masses in children are most commonly represented by neuroblastoma. Non-neuroblastoma tumors (such as adrenocortical carcinomas, pheochromocytomas, teratomas, cysts...) are rare, but early diagnosis is essential due to the risks of malignancy or hormonal hypersecretion.

II. MATERIALS AND METHODS

This is a retrospective descriptive study conducted in the pediatric surgery unit A of the Children's Hospital of Rabat, in collaboration with the pediatric hemato-oncology department. It involved 7 children with adrenal masses other than neuroblastoma, managed over a period of six years between January 2016 and May 2021.

Data were collected from the patients' medical records and included:
- Epidemiological data
- Clinical presentations
- Paraclinical data (imaging and hormonal biology)
- Surgical and therapeutic management
- Postoperative and long-term outcomes

The primary aim was to analyze diagnostic challenges and highlight the role of surgery in managing these rare tumors.

III. RESULTS

Among the 7 patients included:
- 5 had adrenocortical carcinoma
- 2 had pheochromocytoma

The mean age was 6 years (range: 7 months to 14 years), with a sex ratio of 4 females to 3 males.

Clinical presentation included:
- Virilization or precocious puberty in adrenocortical carcinoma cases
- Headache, tachycardia, and arterial hypertension in pheochromocytoma cases Imaging (ultrasound, CT, sometimes MRI) contributed to the diagnosis, supported by hormonal assays (androgens, cortisol, urinary metanephrines).

All patients underwent adrenalectomy. Adjuvant chemotherapy was administered for malignant adrenocortical carcinomas.

The outcomes were favorable in 6 patients, with one case of suspected local recurrence.

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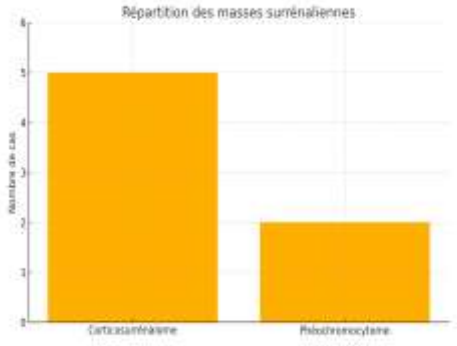

Figure 1. Distribution of adrenal masses in the study population: Among the 7 patients, 5 were diagnosed with adrenocortical carcinoma and 2 with pheochromocytoma.

IV. DISCUSSION

Adrenal tumors in children, excluding neuroblastoma, represent a rare but clinically significant entity. In our series, adrenocortical carcinomas were the most frequent, followed by pheochromocytomas. This aligns with literature data identifying adrenocortical carcinoma as the most common malignant adrenal cortex tumor in children, with an estimated incidence of 0.3 to 0.5 cases per million children per year [1].

Adrenocortical carcinoma mainly affects children under the age of 5, with a female predominance before age 3 [1]. It can be sporadic or associated with genetic syndromes such as Li-Fraumeni syndrome (TP53 mutation), Beckwith-Wiedemann, or Carney complex [2]. Clinical presentation is often dominated by hormonal hypersecretion, particularly androgenic, as observed in our series.

Diagnosis relies on imaging (CT, MRI) and hormonal assessments, but definitive diagnosis is histological [3]. The Weiss score adapted for pediatric use remains a reference for determining malignancy potential in adrenocortical carcinoma.

Treatment is based on complete surgical excision (R0 resection), sometimes followed by adjuvant chemotherapy in proven malignant cases. Laparotomy is generally preferred to avoid tumor rupture, although some studies report satisfactory outcomes with laparoscopic approaches in early-stage tumors under strict conditions [4].

Pheochromocytoma is less common, with an incidence of 0.1 to 0.6 per million children per year. It tends to occur in older children and is often bilateral or familial [5]. It may be isolated or associated with multiple endocrine neoplasia (MEN type 2). Typical symptoms include headache, tachycardia, paroxysmal hypertension, and excessive sweating.

Diagnosis combines imaging and urinary metanephrine and catecholamine assays. Surgery is the treatment of choice, after alpha-blocker preparation to stabilize the hemodynamic status [6]. Laparoscopic approach is possible if the mass is well-circumscribed and without extension.

The prognosis of adrenocortical carcinoma depends on complete resection, absence of metastases, and response to adjuvant treatment. Recurrences often occur within the first two years. Rigorous hormonal and radiological follow-up is essential [7].

Pheochromocytoma also carries a significant risk of local recurrence and contralateral tumor development. Lifelong follow-up is recommended, including for family members in hereditary forms [7].

V. CONCLUSION

Non-neuroblastoma adrenal masses in children are rare and require a multidisciplinary diagnostic and therapeutic approach. Surgical excision remains the cornerstone of treatment. Long-term follow-up is essential to detect recurrence and manage potential late complications. Centralized documentation of these rare cases would help refine epidemiological knowledge and therapeutic strategies.

Figures

Figure 2: CT scan with contrast in a 13-year-old boy showing sagittal (A) and axial (B) views of a hypoechoic tissue-density adrenal mass with heterogeneous enhancement, central necrosis, and some calcifications, suggestive of adrenocortical carcinoma or pheochromocytoma.

A B

Figure 3: Abdominal CT scan with contrast in a 13-month-old girl showing a well-defined, heterogeneous adrenal mass with cystic areas, suggesting adrenocortical carcinoma.

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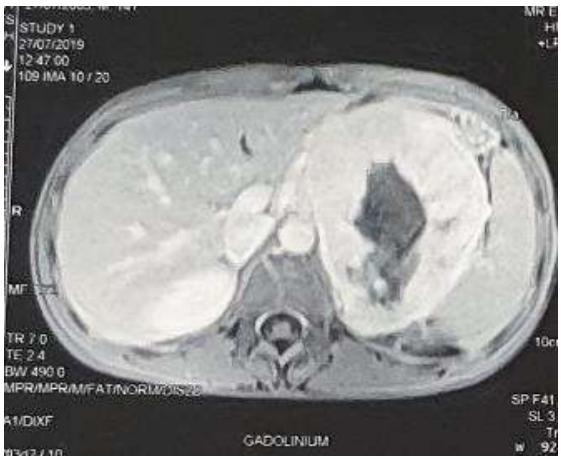
Figure 4: Abdominal MRI – T2 (A) and T1 with gadolinium (B) – showing a left suprarenal retroperitoneal mass with hemorrhagic zones and central necrosis, suggestive of ganglioneuroma.



A

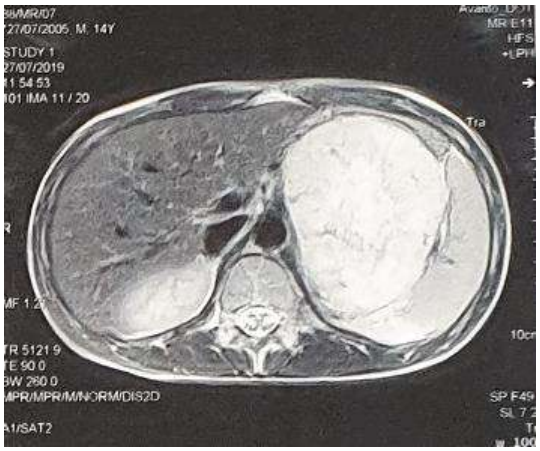


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Figure 5: Axial CT scan in a 4-year-old girl showing a well-defined, oval left adrenal mass measuring 24×18×25 mm.

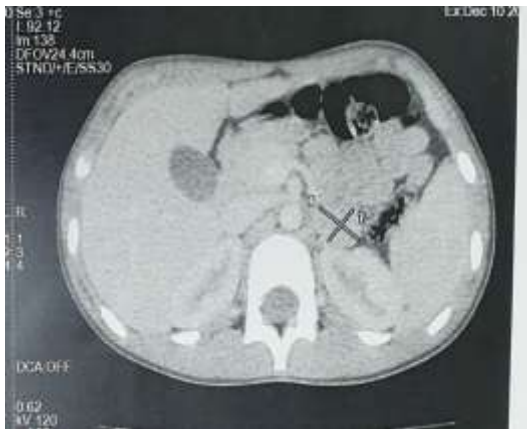
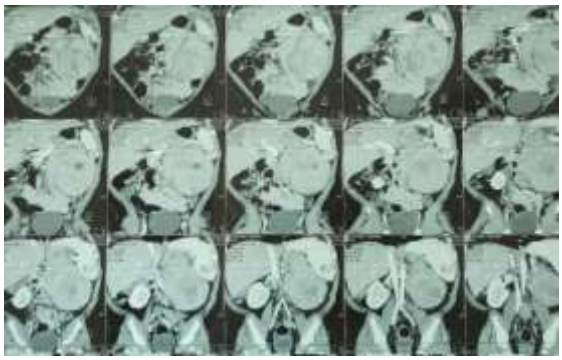


Figure 6: Sagittal (A) and axial (B) CT scans in a 7-month-old boy showing two well-defined left retroperitoneal masses.



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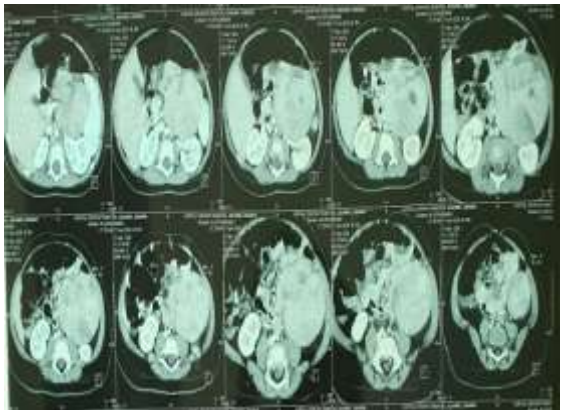

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Figure 7: Sagittal (A) and axial (B) CT scans in a 16-month-old boy showing a well-limited right retroperitoneal tissue mass with heterogeneous enhancement and no calcification.

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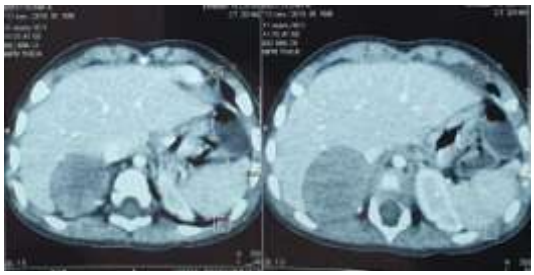

B

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