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Renal Ganglioneuroblastoma in Children: A Case Report

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Abstract

Ganglioneuroblastoma is a neuroblastoma with variable differentiation, surrounded by areas of differentiated ganglioneuroma. It is less common than neuroblastoma and has a better prognosis. We report the case of an 8-year-old boy admitted for the management of a localized retroperitoneal mass. Initial surgery revealed a renal mass, which was completely excised. Histological examination of the surgical specimen confirmed a ganglioneuroblastoma with exclusive renal development-an extremely rare occurrence, highlighting the significance of sharing this case.

Introduction

The Pediatric Oncology Group (POG) identifies three types of tumors forming an anatomopathological continuum: ganglioneuroma, ganglioneuroblastoma, and neuroblastoma [1]. Their prognosis varies based on the tumor's composition of mature and immature cells, degree of differentiation, presence of metastases at diagnosis, the child's age, tumor location, and N-MYC proliferation. These tumors are typically located along the sympathetic chain ganglia and adrenal glands. Tumors originating from the neural crest account for 10% of pediatric cancers in children under 15 years old, with an estimated incidence of 11 cases per million children for ganglioneuroblastoma [2,3]. We report a case of renal ganglioneuroblastoma in an 8-year-old boy, an exceptional location for this type of tumor.

Clinical observation

A 10-year-old male, the eldest of two siblings, has a family history notable for a paternal grandfather who passed away from nasopharyngeal carcinoma, a paternal aunt under follow-up for breast cancer, and a paternal cousin currently being treated for a brain tumor. The patient has been followed since the age of 3 for Autism Spectrum Disorder (ASD). He presented with behavioral disturbances, including sudden agitation episodes, increased autistic withdrawal, and pallor, which prompted the consultation.

Clinical examination revealed a pale patient with growth retardation, afebrile, with normal blood pressure for his height. There was no hematuria or skin abnormalities. The patient exhibited scoliosis and lumbar tenderness, while the remainder of the examination was unremarkable.

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Imaging studies included a total spine CT scan performed to evaluate vertebral malformations, which revealed right-sided scoliosis due to hemivertebrae at L1-L2. Incidentally, a large right retroperitoneal mass was discovered. An abdominal MRI demonstrated a large tumor arising from the right kidney, measuring 11 x 7 x 5 cm, with a bilobed structure, polycyclic contours, hypointensity on T1-weighted images, and tissue-like signal intensity on T2. The tumor showed progressive enhancement with contrast in the excretory phase (Figures 1 and 2).

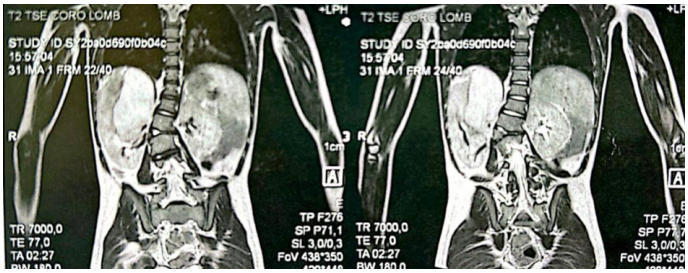

Figure 1: Sociodemographic characteristics of pregnant women in Cotonou.

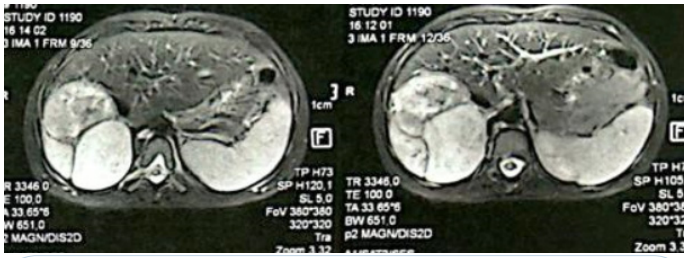

Figure 2: MRI appearance of the renal mass in an axial T2-weighted view.

For tumors involving the kidney, neuroblastoma, or a soft tissue sarcoma, differential diagnoses included nephroblastoma, neuroblastoma, or soft tissue sarcoma.

Biological tests were within normal limits, and tumor markers (blood and urinary catecholamines) were also normal. The remainder of the metastatic workup showed no secondary lesions.

Due to the failure of a radiologically guided biopsy, the case was discussed in a multidisciplinary meeting, a right radical nephrectomy was subsequently performed.

A histopathological study of the surgical specimen revealed macroscopically three nodular masses attached to the kidney: one in the upper pole measuring 10 x 7 x 4 cm, and two parahilar masses, the largest measuring 9 x 7 x 4 cm and the smallest 7 x 5 x 2 cm. On sectioning, the renal mass appeared whitish-yellow, multinodular, firm in consistency, encapsulated, poorly defined, with areas of remodeling and infiltration of the hilar region and sinuses (Figure 3).

Microscopic examination revealed a tumor with a dual component. The first consisted of a ganglioneuromatous proliferation with a predominantly mature Schwannian stroma and mature ganglion cells. The second component was neuroblastic, comprising neuroblastic micronodules with neuroblast cells at various stages of maturation on a neuropil background, accompanied by numerous calcifications. The mitosis-karyorrhexis index (MKI) was low, consistent with the appearance of a nodular ganglioneuroblastoma.

Biopsy of the pararenal lymph nodes showed no evidence of malignancy. In situ hybridization with immunofluorescence of the tumor tissue revealed no N-MYC amplification.

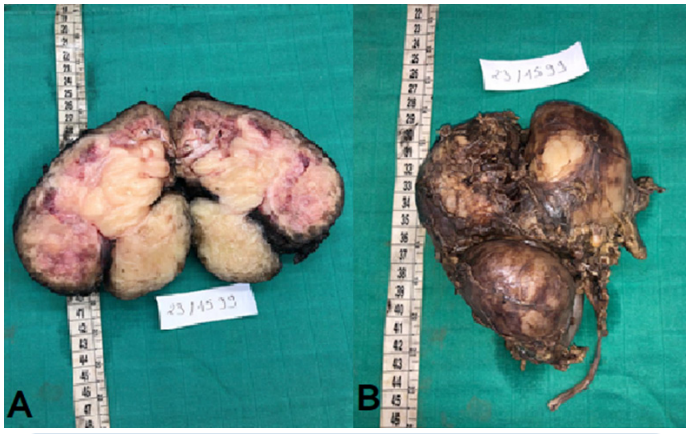

Figure 3: A: Macroscopic section view showing the three masses attached to the kidney.
B: Macroscopic view of the intact surgical specimen revealing a trilobed kidney.

Based on the staging evaluation and prognostic factor analysis (bone marrow aspirates from both iliac crests and a bone marrow biopsy showed no neuroblastic tumor cells; ferritin: 34 ng/mL; Lactate Dehydrogenase (LDH) level: 232 U/L), the patient was classified as low risk.

Regular clinical and radiological follow-up over an 18-month period has shown no evidence of tumor recurrence.

Discussion

We report the case of a 10-year-old patient in whom a renal ganglioneuroblastoma was incidentally discovered during the evaluation of scoliosis.

Renal tumors account for slightly more than 5% of pediatric cancers in Morocco, with a predominance of Wilms tumors. These are often sporadic but can be associated with certain syndromic conditions, such as:

WAGR syndrome (Wilms tumor, aniridia, genitourinary malformations, intellectual disability)

Denys-Drash syndrome (pseudohermaphroditism, glomerulonephritis, Wilms tumor).

Beckwith-Wiedemann syndrome (macroglossia, omphalocele, visceromegaly).

Perlman syndrome (nephroblastomatosis, fetal ascites, macrosomia, Wilms tumor).

Simpson-Golabi-Behmel syndrome (gigantism, intellectual disability, craniofacial anomalies).

They can also occur in the context of genetic instability syndromes, such as Bloom syndrome, Li-Fraumeni syndrome, and incontinentia pigmenti.

Other primary renal tumors in children include:

Clear cell sarcomas of the kidney

Rhabdoid tumors

Renal carcinomas

Mesoblastic nephroma [2,3].

In the study by Nelson et al., conducted over a four-year period from 2004 to 2018 [4], the most common primary site for ganglioneuroblastoma was the thorax, in 40.5% of cases,

followed by the abdomen in 35.7%, specifically at the adrenal medulla and retroperitoneum, but sometimes in the neck and pelvis. No published study reports a renal localization of ganglioneuroblastoma in children.

In the literature, the first case of renal ganglioneuroblastoma was reported in 1990 by Jalleh et al. in England, in a 68-year-old patient who presented with a painless lumbar mass that had been evolving for three weeks without associated renal abnormalities. Histology after nephrectomy confirmed the diagnosis [5]. This tumor is generally silent, with nonspecific clinical manifestations.

A minority of patients, 14%, had distant metastases at the time of diagnosis [3]. In other studies, atypical metastatic sites have been described, such as the thymus, lungs, kidneys, anterior mediastinum, stomach, and cauda equina [6-8].

Ganglioneuroblastoma tends to affect older children, with a median age of 4 years compared to 22 months for neuroblastoma [9]. The average age at diagnosis is 47.5 months, with 90% of cases diagnosed before the age of 5. Tumors diagnosed after the age of 10 are extremely rare [10]. In our patient, the ganglioneuroblastoma was discovered at the age of 10.

Radiology helps assess the mass, its relationships, and establish an extent evaluation, but only histology combined with molecular biology can confirm the diagnosis and establish the prognosis. Abdominal CT is the imaging modality of choice for evaluating neuroblastic tumors; the mass is well-defined, heterogeneous, with the presence of calcifications in 50% of cases, and sometimes necrotic areas [11]. On abdominal MRI, the tumor appears heterogeneous with hypointensity on T1 and tissue signal on T2 [11]. In our case, MRI shows a large retroperitoneal tumor process with heterogeneous T2 signal due to the renal excretion time of gadolinium.

Ganglioneuroblastomas are histologically composed of two components: the malignant neuroblastic component, whose differentiation and proliferation characteristics, particularly the Mitosis-Karyorrhexis Index (MKI), must be studied, along with N-MYC amplification to determine the therapeutic risk group. The benign component is represented by a differentiated ganglioneuroma.

The INPC (International Neuroblastic Pathology Committee) defines two types of ganglioneuroblastoma [12]: mixed ganglioneuroblastoma, with a rich and poor composite stroma: it is primarily composed of Schwann cells, which represent more than 50% of the entire tumor, with the presence of small clusters or masses of 10 to 30 tumor cells on average, sometimes more, showing varying differentiation but often with ganglion-like tumor cells scattered in a well-defined fibrillar background. The second type is nodular ganglioneuroblastoma, with a rich and poor composite stroma, which can be either classic (single nodule) or variant, with either multiple nodules or large nodules, single or multiple. The macroscopically visible nodule is often hemorrhagic and has a sharp boundary with the non-nodular component of the tumor, which is histologically either ganglioneuroma or mixed ganglioneuroblastoma. In the nodule, the tumor is exclusively composed of neuroblasts at various stages of maturation, which can thus be undifferentiated or poorly differentiated, and more rarely, differentiating. Our patient had a renal nodular ganglioneuroblastoma.

The treatment is surgical resection with or without chemotherapy and/or radiotherapy, depending on the histological risk group of the neuroblastic component, N-MYC amplification, and the presence or absence of metastases [6]. In the literature, the majority of patients have been treated with surgical resection without adjuvant therapy [12,13].

Conclusion

Ganglioneuroblastoma is a rare tumor in children. Renal localization is exceptional, and no cases of renal ganglioneuroblastoma in children have been described in the literature. It generally has a good prognosis if diagnosed early and correctly, highlighting the importance of this publication. Management primarily relies on complete surgical resection in localized forms. The presence of distant metastases at the time of diagnosis is rare and may worsen the prognosis.

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