CLINICAL CASE

BILATERAL NEPHROBLASTOMA – CASE PRESENTATION

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Abstract

Nephroblastoma is the most common pediatric abdominal malignancy, representing approximately 30% of pediatric cancers.Our case concerns a 3-year-old girl diagnosed with bilateral nephroblastoma, who presented 4 renal tumors, of which one that originated in the left kidney was occupying almost entirely the left half of her abdomen. The patient, D. A., was hospitalized in the surgery department of „Maria Sklodowska Curie" Emergency Children's Hospital in Bucharest on 10.10.2014 in order to continue investigations regarding a right renal mass discovered incidentally during a routine abdominal echography. An abdominal CT scan revealed 2 masses in her left kidney, the largest of which measured 12/8/9 cm, and another 2 in her right kidney.Clinical and paraclinical data was suggestive for nephroblastoma, so a 4 week combined chemotherapy regimen (Vincristine and Actinomycin D) was initiated. A second abdominal CT showed a significant reduction in the size of the masses, which prompted physicians to repeat the regimen. Afterwards, a third abdominal CT scan showed an even further reduction in size, the largest mass now measuring 49/43 mm.The patient underwent a left ureteronephrectomy on 24.12.2014, along with the excision of the 2 right renal tumors. Postoperatively, a third chemotherapy regimen was initiated, with the same medication, this time lasting 27 weeks. At the time of publishing, the patient is still under chemotherapy treatment. This case presents a few interesting features, highlighting the possibility of a very positive response to treatment of nephroblastoma, despite a dire initial prognosis.

Keywords*: nephroblastoma, bilateral, child, chemotherapy, ureteronephrectomy*

Introduction

Nephroblastoma, also known as Wilms tumor, is a renal malignancy which presents a few very interesting specific features, including a high incidence rate as well as a high rate of treatability/curability, in comparison to other pediatric cancers.

It is the most common pediatric renal malignancy, representing 85% of all pediatric renal cancers [1] and 6% of all pediatric malignancies [2]. The average age at diagnosis is 44 months in case of unilateral presentation and 31 months in case of bilateral presentation [3,4]. Approximately 10% of patients present associated congenital malformations [5].

Survival rates for patients vary from 33% to 98% and are dependent on the following factors: staging at diagnosis (Table 1), size of mass, patient age and histopathological characteristics (survival rate decreases as degree of anaplasia increases) [7-10].

Nephroblastoma treatment can be surgery, chemotherapy or radiotherapy, and is individualised according to staging and aggresiveness. Surgical treatment consists of

nephrectomy associated with lymphadenectomy. Radiotherapy is indicated only in advanced stages [10], whilst chemotherapy plays a decisive role and is indicated in all stages, properties such as regimen intensity, frequency and length being established according to prognosis.

Stage	Characteristics
I	The tumor is exclusively renal and resectable
II	Tumor invasion beyond the kidney but not out of the renal area, still being resectable
III	The tumor has spread into the abdominal cavity, with invasion into the lymph nodes, viscera or neighboring vessels
IV	Tumor metastases in bone marrow, lungs, brain or liver
V	Bilateral tumors

Table 1 - Nephroblastoma staging

Case presentation

D. A., aged 38 months, was hospitalized in the surgery department of „Maria Sklodowska Curie" Emergency Children's Hospital in Bucharest on 10.10.2014 in order to continue investigations regarding a right renal mass discovered incidentally during a routine abdominal echography.

Examination revealed an otherwise healthy child with a large mass occupying almost the entire left half of the abdomen, along with slight pain upon palpation of the right kidney. Giordano's sign is negative bilaterally.

Initial blood work revealed a slight degree of anemia, along with an inflammatory reaction (Table 2). Homovanillic acid and vanillylmandelic acid levels were normal.

Index	Value	Normal limits
Hemoglobin	10.3 g/dL	≥ 11.5 g/dL [12]
Hematocrit	31.2%	36-40% [13]
ESR	120 mm/h	≤ 20 mm/h [14]
CRP	18.73 mg/dL	≤ 1 mg/dL [15]

Table 2 - Selected abnormal results of initial blood work.

The patient was diagnosed with bilateral nephroblastoma.

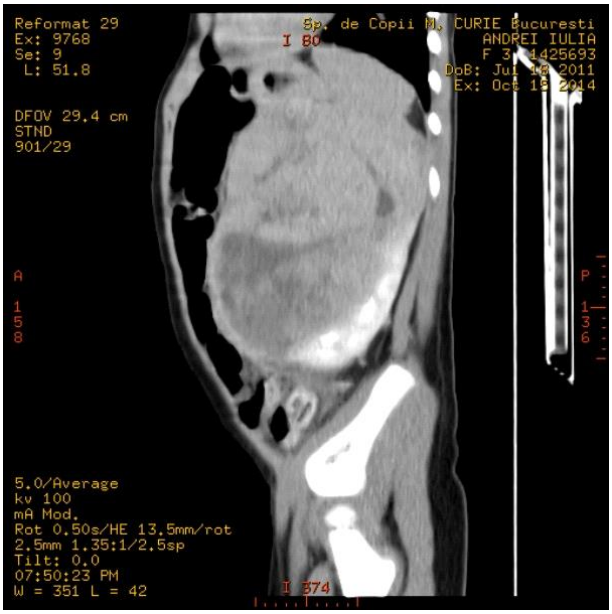

Figure 1 - Tumor of the left kidney at the superior pole, measuring 12/8/9 cm and occupying almost ¾ of the renal parenchyma and almost the entire left half of the abdomen

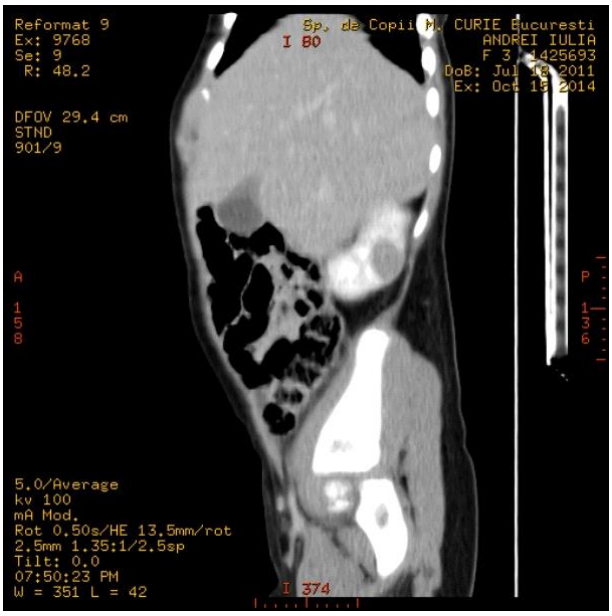

Figure 2 - One of the two right renal rumors

An abdominal CT scan showed modified architecture of the left kidney, which presents 2 masses. The larger one, measuring 12/8/9 cm, had well defined boundaries and occupied ¾ of the renal parenchyma, almost filling the left half of the abdomen and displacing the spleen cranially (Figures 1 and 4). The second, situated at the inferior pole and extending medially, measured 5/4/5 cm. Both presented heterogenous structure and were destroying the

renal parenchyma. The right kidney also presented 2 masses near the inferior pole, measuring 3/2.5 cm and 2/1.5 cm respectively. Their boundaries were clearly defined and they presented homogenous structure (Figures 2, 3 and 4). A lomboaortic lymphadenopathy was also observed. No other abdominal lesions or anomalies were reported.

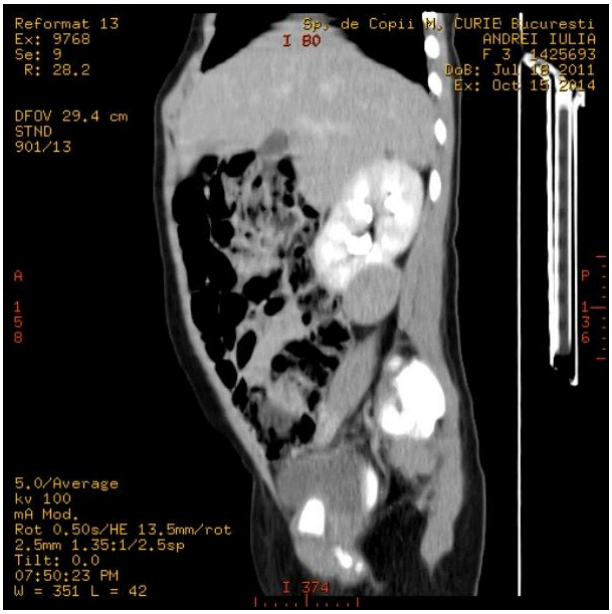

Figure 3 - The second right renal mass

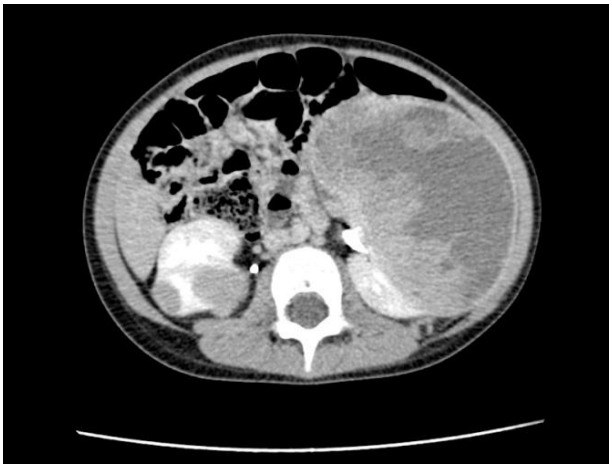
Figure 4 - Cross section that simultaneously captures three of the four masses (2 in the right kidney and the larger of the 2 in the left kidney)

Material and Methods

On 20.10.2014, the first regimen of chemotherapy was initiated, according to the 2001 SIOP protocol for bilateral nephroblastoma (Table 3).

Week	Medication
I (20.10)	Vincristine 1 mg Cosmegen (Actinomycin D) 700 µg
II (27.10)	Vincristine 1 mg
III (03.11)	Vincristine 0.9 mg Cosmegen (Actinomycin D) 675 µg
IV (10.11)	Vincristine 1 mg

Table 3 - The first chemotherapy regimen

Abdominal CT scan performed on 12.11.2014 showed a significant size reduction of all 4 renal masses, all of which now presented areas of necrosis. The 2 left renal masses now measured 71/57 mm and 25/18 mm respectively, whilst the 2 right renal masses measured 18/15 mm and 19/13 mm respectively.

The patient underwent a second chemotherapy regimen, in accordance with protocol (Table 4).

Week	Medication
I (17.11)	Vincristine 1 mg Cosmegen (Actinomycin D) 675 µg
II (24.11)	Vincristine 1 mg
III (04.12)	Vincristine 1 mg Cosmegen (Actinomycin D) 675 µg
IV (11.12)	Vincristine 1 mg

Table 4 - The second chemotherapy regimen.

Abdominal CT scan performed on 17.12.2014 revealed further improvement, the 2 left renal masses now measuring 49/43 mm and 29/28 mm respectively (Figure 5), and the 2 right renal masses 17/16 mm and 6/5 mm respectively (Figure 6). Infracentrimetric retroperitoneal lymphadenopathies were also noted.

The patient underwent surgery on 24.12.2014. Surgery consisted in left ureteronephrectomy, along with excision of left perihilar and paraaortic lymph nodes, which were hardened and increased in volume, as well as excision of the 2 right renal masses (Figure 7), along with the excision of right perihilar and pericaval lymph nodes and one mesenteric lymph node, all increased in size and hardened.

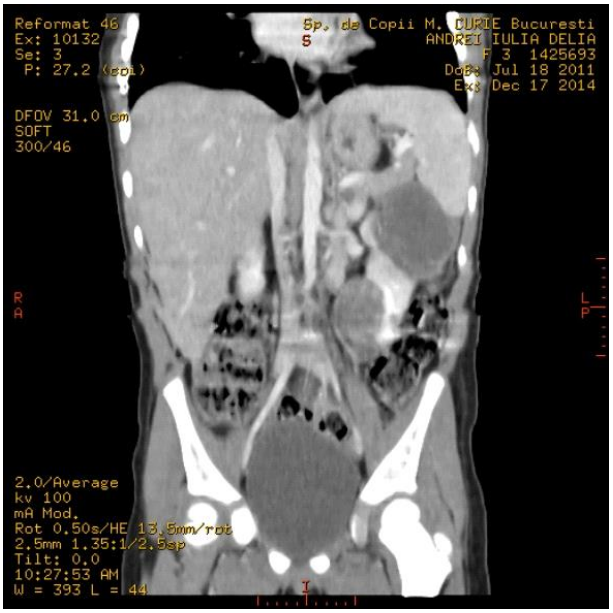

Figure 5 - Significant decrease in size of the left renal tumors after the 2 rounds of chemotherapy

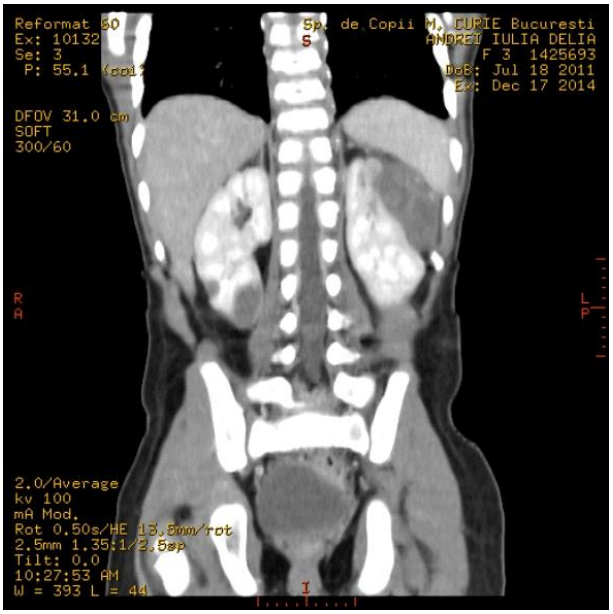

Figure 6. The right renal tumors also decreased in size after the 2 rounds of chemotherapy

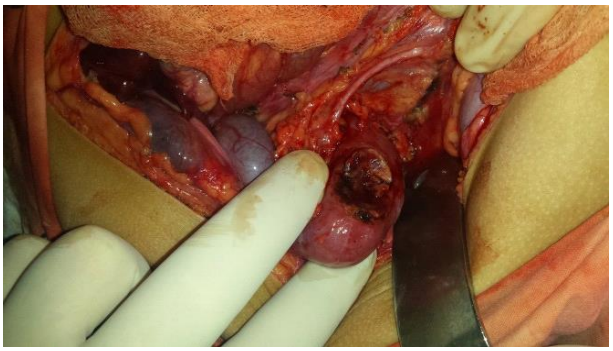

Figure 7 - Dissection of one of the tumors from the right kidney

The histopathological diagnosis was bilateral nephroblastoma (SIOP stage 5). All of the excised lymph nodes presented only sinus histiocytosis.

Postoperative chemotherapy is initiated on 09.01.2015 (Figure 8).

The patient is currently undergoing this final chemotherapy regimen.

Figure 8 - Postoperative chemotherapy regimen, administered according to the same 2001 SIOP protocol

Discussions

This case presents several interesting unique features, the first of which being that the patient presented 4 independent renal masses. Also, given the advanced stage of the disease upon diagnosis, the lack of symptoms and the presence of only a few clinical signs was quite surprising.

It also serves to highlight the importance of good cooperation and communication among members of the treatment team in oncological cases, virtues that make therapeutic success in such cases a real possibility.

Long-term follow-up investigations may prove interesting as well, as this child currently has a very questionable prognosis regarding both recurrence of the cancer and complications due to the reduced remaining renal volume and function.

Detail has been removed from this case description to ensure anonymity. The editors and reviewers have seen the detailed information available and are satisfied that the information backs up the case the authors are making.

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