



Case Series: Rhabdoid Kidney Tumor and Literature Review

Uribe RK^{1*}, Ruiz AN², Morante DC³, Cruz R⁴, Hernández E⁵, García LJ⁶, García EM⁶ and Destefano UV³

¹Department of Surgical Oncology, National Cancer Institute (INEN), Peru

²Urology Oncology Fellow, National Cancer Institute (INEN), Peru

³Department of Urological Surgery, National Cancer Institute (INEN), Peru

⁴Pathology Oncology Fellow, National Cancer Institute (INEN), Peru

⁵Department of Pediatric Oncology, National Cancer Institute (INEN), Peru

⁶Department of Clinical Oncology, Regional Hospital of Lambayeque, Peru

Abstract

Background: Rhabdoid kidney tumors are a rare infant entity. We present a case series of 10 patients with this diagnosis in a period of 19 years at the National Cancer Institute of Peru.

Methods and Results: Descriptive and retrospective study. We analyzed patients with rhabdoid kidney tumor in the Department of Urological Surgery of the National Cancer Institute (Instituto Nacional de Enfermedades Neoplásicas: INEN) in Lima-Peru, from January 2000 to December 2019. The results: 10 cases were reported with anatomopathological diagnosis of Rhabdoid kidney tumor.

According to clinical characteristics: Seven male patients (70%) and three females (30%), the proportion of population related to age group was: Seven infants (between 1 to 23 months) (70%), two preschool (between 24 to 71 months) (20%), one child (between 6 to 12 years) (10%). About symptoms six patients present hematuria (60%), three patients with palpable tumor mass (30%) and one patient with fever (10%). Paraneoplastic hypercalcemia was infrequent with two cases (20%).

According to surgical characteristics: Left kidney was the most frequent presentation in six cases (60%). Average tumor size was 10 cm. Of the ten patients, surgical treatment was made in nine patients (90%), of which seven (70%) had primary surgery, two (20%) had surgery post neoadjuvant treatment and one case did not have surgical treatment (due to progressive disease). R0 in seven cases (70%) and R1 in two cases (20%).

Related to multimodal treatment, at the chemotherapy arm, eight patients (80%) received systemic chemotherapy; three of them (30%) had neoadjuvant therapy, two of them had complementary surgery (of these two patients, one had complementary adjuvant chemotherapy and the other one surgical management post neoadjuvant treatment without adjuvant treatment); the other one patient did not have surgical management in relation of progressive disease. The remaining five patients (50%) had adjuvant treatment posterior primary surgical management. One patient had adjuvant radiotherapy (10%) and one patient (10%) had surgical management without complementary treatment. Prevalent clinical stage was III in five cases (50%), and medial global survival of seven months.

Conclusion: This entity is predominantly in young children, most of cases with advanced stage disease and poor survival even if the diagnosis is on early stage.

Keywords: Rhabdoid kidney tumor; Children kidney cancer; Radical nephrectomy

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*Correspondence:

Karla Uribe Rivera, Department of Surgical Oncology, National Cancer Institute (INEN), Lima, Peru,
E-mail: karla.uriberivera@gmail.com

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Introduction

Kidney cancer in children represents the 7% of all new cases of pediatric cancer in the United States, the registration of Malignant Rhabdoid Tumor of the Kidney (MRTK) accounts 158 (1.6%) of 10,031 registrants' renal tumors [1]. The most common histology for kidney cancer in children is Wilms tumor (75% of cases) with a high overall survival rate (>90%). Other variants with poor prognosis are the Anaplastic Histology of Wilms Tumor (AHWT) and Malignant Rhabdoid Tumor (MRT), the last one is known to have among the worst prognosis for solid tumors of childhood,

without report of incidence in most countries [2].

Between 1984 and 1999, approximately six patients per year diagnosed with MRTK were enrolled on varied protocols in Germany. Between 1993 and 2005, 207 MRTK patients were enrolled on the renal tumor treatment study group protocols of The International Society of Pediatric Oncology (SIOP). The Children's Oncology Group (COG) and its predecessor, the National Wilms Tumor Study Group (NWTSG) along with the SIOP, have been responsible for multiple trials over the last 40 years, with significant advances in rhabdoid tumors which are highly malignant, originating from any organ with worst prognosis for childhood solid tumors.

In our country, the last data of cancer in Metropolitan Lima was publishing in 2012 and the incidence of cancer in pediatric population was 1.7% of all the cases. Actual information related to renal cancer incidence between 2000-2018 in the National Cancer Institute (INEN) statistics register a median of 206 new cases per year [3].

The goal of this study is to contribute to the global statistics of this rare entity, increasing studies and to develop multidisciplinary management.

Materials and Methods

This is a descriptive and retrospective study. We analyzed patients with rhabdoid kidney tumor in the Department of Urological Surgery of the National Cancer Institute (Instituto Nacional de Enfermedades Neoplásicas: INEN) in Lima-Peru, from January 2000 to December 2019. Patients with anatomopathological confirmation of Rhabdoid tumor of the kidney that had treatment with surgery, chemotherapy and/or radiotherapy were followed in this period.

The clinical characteristics were: Gender, age of presentation, principal symptoms and paraneoplastic syndromes associated. The surgical characteristics were: Tumor location and size, chemo/radiotherapy association, type of surgery (debut, post neoadjuvant treatment), type of procedure, kind of resection (R0-R1-R2), lymph node commit, clinical stage, overall survival and progressive disease.

As a case series, we only present absolute numbers and frequencies data, which are presented in the statistical part.

The proportion of cases diagnosed with solid renal tumors in our institution, are between 200 to 300 cases per year, and rhabdoid kidney tumors are approximately 1 case per year, which represents the 1.6% of our population in this last eighteen years.

Results

We got 10 cases in a period of 19 years, with pathological

confirmation of rhabdoid tumor of the kidney. According clinical characteristics: Seven patients' male (70%) and three females (30%), the proportion of population related to age group: Seven infants (between 1 to 23 months) (70%), two preschool (between 24 to 71 months) (20%), one child (between 6 to 12 years) (10%). About symptoms six patients present hematuria (60%), three patients (30%) with palpable tumor mass, two of these cases associated with abdominal pain and one patient with fever (10%). Paraneoplastic hypercalcemia was infrequent with only two cases (20%) (Table 1).

According to surgical characteristics: Predominantly presentation was left kidney in six cases (60%), right kidney in three cases (30%) and bilateral in one case (10%). The average tumor size was 10 cm. Of the ten patients, surgical treatment was made in nine patients (90%), of which seven (70%) had primary surgery, three (30%) had neoadjuvant treatment and one of these cases did not have surgical treatment (due to progressive disease). R0 in seven cases (70%) and R1 in two cases (20%).

Related to multimodal treatment, in the chemotherapy arm, eight patients (80%) received systemic chemotherapy; three of them (30%) had neoadjuvant therapy, two of them had complementary surgery (of these two patients, one had complementary adjuvant chemotherapy and the other one had surgical management post neoadjuvant treatment without complementary adjuvant); the other one patient did not have surgical management in relation of progressive disease.

The remaining five patients (50%) had adjuvant treatment posterior primary surgical management. One patient had adjuvant radiotherapy (10%) and one patient (10%) had surgical management without complementary treatment.

Clinical stage was: I none, II in 2 cases (20%), III in 5 cases (50%), IV in 2 cases (20%), V in one case (10%), progressive disease in 6 cases (60%) with medial global survival of 7 months (Table 2).

Discussion

Originally, rhabdoid kidney tumor thought to be a variant of Wilms tumor, until 1978 when it was designated as a separate etiology. They are aggressive cancers presenting in infants and young children, 60% occurring before 1 year of age, and 80% before 2 years with a median age of 10.6 months (mean 15 months) [2].

According to the International Society of Pediatric Oncology (SIOP), their incidence is 0.9% of pediatric kidney tumors [4] and 1.8% of malignant childhood kidney tumors in the National Study of Wilms tumors. In our institutional database, 532 pediatric patients

Table 1: Clinical characteristics.

Item	Gender	Age (m)	Main Symptom	Paraneoplastic Syndrome
1	M	6	Hematuria	No
2	M	2	Hematuria	No
3	M	10	Hematuria	Hypercalcemia
4	M	9	Hematuria	No
5	F	12	Fever	Hypercalcemia, hypokalemia
6	M	72	Abdominal pain and palpable tumor mass	No
7	M	24	Abdominal pain and palpable tumor mass	No
8	M	9	Hematuria	No
9	F	12	Abdominal pain	No
10	F	48	Hematuria	No

Table 2: Surgical characteristics.

Item	Location	Tumor size (cm) Pre-Chemo-Therapy Ct Scan	Tumor size (cm) post chemo-therapy - CT scan	Chemotherapy	Radiotherapy	Protocol	Type of chemotherapy	Debut surgery	Procedure	Resection	Lymph node. Commit	Stage	OS (m)	Progression
1	Right	7.5	NO NEO-ADJUVANT	ADJUVANT CHEMOTHERAPY	NO	PROTOCOL 9229	ACTINOMYCIN-D and VINCRISTINE	YES	RIGHT RADICAL NEPHRECTOMY	R0	0/0	III	5	INTRA TORAXIC TUMOR
2	Bilateral	R: 9 L: 4.5	R: 11 L: 4.5	NEO-ADJUVANT CHEMOTHERAPY	NO	PROTOCOL 9229	DOXORUBICIN - ACTINOMYCIN - VINCRISTINE	NO	RIGHT RADICAL NEPHRECTOMY	R0	0/1	V	one	LOCAL PERSISTENCE
3	Left	10	NO NEOADJ	ADJUVANT CHEMOTHERAPY	NO	MEM 0321	DOXORUBICIN	YES	LEFT RADICAL NEPHRECTOMY	R0	0/0	IV	two	NO REGISTRATION
4	Left	20	NO NEOADJ	ADJUVANT CHEMOTHERAPY	NO	INDIVIDUAL PROTOCOL – ACCORDING TO COG	CYCLOPHOSPHAMIDE - DOXORUBICIN - CARBOPLATIN	YES	LEFT RADICAL NEPHRECTOMY + FOR AORTIC LIMPHADENECTOMY	R0	0/2 RENAL HILUM + 0/4 FOR AORTIC	II	84	NOT
5	Right	8	NO NEOADJ	ADJUVANT CHEMOTHERAPY	NO	INDIVIDUAL PROTOCOL- ACCORDING TO COG	CYCLOPHOSPHAMIDE - CARBOPLATIN - ETOPOSIDE	YES	RIGHT RADICAL NEPHRECTOMY	R0	1/1 PARACAVAL + 1/1 RETROCAVAL	II	19	LOCAL PERSISTENCE, LUNG AND BONE PROGRESSION
6	Left	12	NO NEOADJ	NO	NO	NO	NO	YES	LEFT RADICAL NEPHRECTOMY	R0	0/1	III	two	LOCAL PERSISTENCE, BONE PROGRESSION
7	Left	10	NO NEOADJ	ADJUVANT CHEMOTHERAPY	NO	INDIVIDUAL PROTOCOL – ACCORDING TO COG	DOXORUBICIN - CYCLOPHOSPHAMIDE - VINCRISTINE	YES	LEFT RADICAL NEPHRECTOMY + SPLENECTOMY + RETROPERITONEAL LYMPHADENECTOMY	R1	FOR AORTIC/HILUM AND SPLENIC LYMPHADENOPATHY	III	216	NO REGISTRATION
8	Right	12	NO NEOADJ	NO	ADJUVANT RADIO-THERAPY	NO	NO	YES	RIGHT RADICAL NEPHRECTOMY + THROMBECTOMY OF RIGHT AND LEFT RENAL VEIN/ INFERIOR CAVA	R1	0/0. RENAL VEIN THROMBOSIS	III	228	NO REGISTRATION
9	Left	14	8	NEO-ADJUVANT AND ADJUVANT CHEMOTHERAPY	NO	INDIVIDUAL PROTOCOL – ACCORDING TO COG	DOXORUBICIN - CYCLOPHOSPHAMIDE - VINCRISTINE	NO	LEFT RADICAL NEPHRECTOMY	R0	0/0. RENAL VEIN INFILTRATION	III	8	LOCAL RECURRENCE
10	Left	10	9	NEO-ADJUVANT CHEMOTHERAPY	NO	INDIVIDUAL PROTOCOL – ACCORDING TO COG	CYCLOPHOSPHAMIDE - CARBOPLATIN - ETOPOSIDE	NO	NO SURGERY MANAGEMENT	NO	0/0	IV	6	PROGRESIVE DISEASE

were diagnosed with solid renal tumors between 2000-2018 and rhabdoid kidney tumor represents the 1.6% of cases of our population.

In general, it occurs mainly in children younger than 2 years, with a mean age at diagnosis of 11 months, which coincides with our report where we found seven patients younger than 2 years and the average age of the 10 patients was 20 months (age ranges between 2 to 72 months) [4].

Those tumors are extremely rare in adult patients, there are only five cases of adults been reported at English literature and one case in China [5]. In our results we found that 70% male and 30% female; according age groups, 70% were infants, 20% preschool and 10% scholar; male predominance from 2 to 1 [6], median age was 11 months, age range from: 0 to 116 months and a male-female ratio of 1.5 to 1, predominant location in left kidney is according to results of others case series and mean size tumor was 8 cm [7].

The clinical stages in our study were: I: 0% - II: 20% - III: 50% - IV: 20% - V: 10%, comparable to the study by Palmer (6), with clinical stage I (10%), 10 patients clinical stage II (47%), 5 clinical stage III (23%) and 4 patients with clinical stage IV (20%). Also, the predominance of left kidney it's according results of others case series [8].

Abdominal mass and hematuria are common symptom in urogenital cancer, children are not the exception. Paraneoplastic syndromes are related to this entity, it is relevant to mention hypercalcemia that occurs as effect of the increased concentration of parathyroid hormone; in addition the symptoms that arise from metastatic disease are common at the time of diagnosis.

These tumors are typically large, hemorrhagic and necrotic, with

poorly defined borders, characterized by a pale cut surface [8], a feature that reflects their highly invasive nature.

Macroscopically without capsule and highly infiltrative borders [8]. They are composed of sheets of relatively uniform malignant round cells, which aggressively overrun native nephrons, and vascular invasion is often wide. Rhabdoid tumor cells display the triad of vesicular chromatin, prominent cherry-red nucleoli and hyaline pink cytoplasmic inclusions [9]. These globular inclusions correspond ultra structural to whorled intermediate filaments [10]. Nonetheless, the prominence and number of rhabdoid cells may vary from one microscopic field to another and from one tumor to another; several other patterns exist, such as epithelioid, sclerosing, spindle and lymphomatoid [8]. A subset of rhabdoid kidney tumors is composed of predominantly primitive undifferentiated small round cells.

The immunohistochemistry sample shows that tumor cells are diffuse and intense reactive for vimentin, focal reactive for cytokeratin and EMA [8]. Loss of INI-1 (SMARCB1) is a sensitive and specific marker of this pathology [11], although such loss is seen in other neoplasms, including renal medullary carcinoma and epithelioid sarcoma [12-14]. Other typically negative markers for this tumor are NSE, S100, CD99, Desmin and Leu [10].

In the genetic profile, biallelic inactivation of the SMARCB1 tumor suppressor gene, which resides on the long arm of chromosome 22, is the molecular hallmark of rhabdoid tumor of the kidney [15,16]. Inactivation of this gene is also found in tumors with morphologically similar features, in soft tissue and in the brain (called atypical teratoid/ rhabdoid tumors of the brain) and occasionally in other visceral sites. All of these tumors affect young children and are usually lethal.

Children with concurrent rhabdoid tumor of the kidney and

atypical teratoid/rhabdoid tumor of the central nervous system often harbor germline mutations in the SMARCB1 gene [17,18]. The inactivation of the second allele occurs by distinct mechanisms in these two neoplasms, which confirm the clinicopathological impression that these are independent entities [19].

The SMARCB1 gene encodes a protein involved in the SWI/SNF chromatin-remodeling complex, and its inactivation is thought to promote neoplasm by altering gene expression. Inactivation of SMARCB1 take place in mutational *via* deletion or whole chromosome loss, which may explain the common cytogenetic finding of monosomy 22 in these tumors [18].

There are some rhabdoid tumors of the kidney with intact SMARCB1 expression, but these tumors harbor mutations in the SMARCA4/BRG1 gene, which encode a protein of the same complex.

In relation to differential diagnosis, Wilms tumor characteristically has blastemal, epithelial and mesenchymal components. Tumor cells are positive for WT1 and CD56. Occasionally, rhabdoid tumors lack the typical cytoplasmic inclusions and may resemble other small blue cell tumor of the childhood, such as Ewing sarcoma and neuroblastoma. Immunostains including cytokeratin and neuroendocrine markers and CD99 are usually helpful to establish a correct diagnosis [10].

Surgery is considered the first treatment option if it is possible [20], which agrees with our results where the initial surgical treatment was 80% with a pathological result of R0: 70%, R1: 20% and one case did not perform surgery in relation to progressive disease (10%). According to the study by Wagner et al. [7], rhabdoid tumor of the kidney is one of the deadliest malignancies in pediatric oncology; the median age of diagnosis is between 10 to 18 months of age. Metastatic disease comes early in the first two years in contradistinction to nephroblastoma stage IV tumors, where the metastatic disease is rare [21].

Like other pediatric renal tumors, low stage tumors have a better outcome, the age at diagnosis; however, is a previously unrecognized important prognostic factor. Children less than a year of age at diagnosis have bad prognosis, while those older than 1 year of age at diagnosis have a modestly better prognosis.

Case reports and small series suggest chemosensitivity of rhabdoid tumor of the kidney to anthracyclines, alkylating agents, such as platinum derivatives and oxazophosphorines with radiation therapy.

These patients have been treated using different protocols over the last decades [22], the regimens including vincristine, actinomycin D, doxorubicin, and cyclophosphamide have resulted in disease-free survival estimates of only 19% to 25% [23]; the ICE regimens (Ifosfamide, Carboplatin, Etoposide) and VDC (Vincristine, Doxorubicin, Cyclophosphamide) have also been used after not responding to primary treatment with cyclophosphamide, alternating with carboplatin and etoposide. It is unclear which agent or agents provide the greatest benefit, and whether or not ifosfamide has a therapeutic advantage over cyclophosphamide, doxorubicin; or whether the synergy of combinations ICE and VDC explain response. Gururangan et al. [24] reported encouraging but short lasting, responses to ICE in patients with advanced-stage rhabdoid kidney tumor.

In our study, neoadjuvant chemotherapy regimens were used

in 20% of cases, based on vincristine, actinomycin D, doxorubicin or cyclophosphamide, carboplatin, etoposide, adjuvant in 50% with doxorubicin, cyclophosphamide, vincristine or cyclophosphamide, neo, adjuvant, 10% adjuvant, doxoropuvant; radiotherapy in one case (10%), and no chemotherapy in one case (10%). The highest survival was 228 months in which the patient received adjuvant radiotherapy, followed by 216 months who received adjuvant chemotherapy based on doxorubicin, cyclophosphamide, and vincristine, and 84 months with cyclophosphamide, doxorubicin and carboplatin.

Patients younger than 2 years or older than 18 years at diagnosis, have survival rates lower than patients between 2 and 18 years, which coincide with our results, where it was found that the median overall survival of the 10 patients was 7 months, the lowest survival in EC V of 1 month, followed by 2 months in clinical stage III and IV. In addition, the patient who achieved the highest survival of 228 months was clinical stage III and received adjuvant radiotherapy, similar to the study by Chung et al. where the survival time of 8 patients varied from 1 week to 20 months (with an average: 6 months) [21].

The prognosis for children with rhabdoid kidney tumors is poor, which is confirmed in the study by Palmer et al. [6] where only 2 of the 21 patients from 1969 to 1978 survived: Both treated with the NWTs Regimen that combines Adriamycin, vincristine, actinomycin - D, and postoperative irradiation of the tumor bed, compared to our study where we had four live patients.

The diagnosis and management of this entity is a challenge, with poor prognosis; nonetheless, it is necessary to report these cases and improve the knowledge with help of world statistics. More clinical trials are also needed in order to improve management of this rare pathology.

Conclusion

We conclude that this entity is predominantly in young children, most of cases with advanced stage disease, with poor survival even if the diagnosis is on early stage.

Our study is the first in our country that collects data of the multimodal treatment however, the proportion of cases reported in the international literature are few, and further trials are need for conclude management.

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