



NEPHROBLASTOMA: PRESENTATION OF A CLINICAL CASE

Alexánder Antonio Jara Chávez	MD. General Practitioner *Corresponding Author / Quito - Ecuador
Carolina Vanessa Saldaña Rodríguez*	MD. General Practitioner / Quito - Ecuador *Corresponding Author
Carlos Alberto Lara Rueda	Medical resident at "Hospital de las Fuerzas Armadas N1 / Quito – Ecuador
Priscila Monserrate Cisneros Andrade	MD. General Practitioner / Quito – Ecuador
Víctor Alfonso Terán Pérez	Medical resident at "Clinica Pasteur" Quito – Ecuador
Nadya Steffany Reyes Nieto	Medical resident at "Hospital Enrique Garces" / Quito – Ecuador
María Belén Proaño Bonifaz	Medical resident at "Hospital de las Fuerzas Armadas N1 / Quito – Ecuador
Betty Fernandez Santamaria	MD. General Practitioner / Quito - Ecuador

ABSTRACT

Objectives: To describe the frequency, possible causes, symptoms, diagnosis, prognosis, complications and treatment of this disease.

Method: A study of a pediatric patient with a right renal tumor lesion was performed, who underwent a clinical-surgical procedure and post-treatment follow-up.

Result: A 3-year-old girl, who was diagnosed with a right renal tumor, underwent Right Nephroureterectomy + Appendectomy, confirming the diagnosis of Nephroblastoma. Subsequently, he underwent chemotherapy treatment. Patient 4 years after the end of treatment is free of disease.

Conclusion: Early and timely detection together with a correct tumor staging and adequate multidisciplinary treatment is the objective to achieve control of the disease, even achieving cure.

KEYWORDS : Nephroblastoma, nephroureterectomy, Quito, Ecuador

INTRODUCTION

Nephroblastoma or Wilms' tumor. Malignant neoplasm of the kidney, the second most common type of pediatric abdominal cancer.

It typically debuts in childhood (1 in 200,000 to 250,000 children), uncommon in newborns and ages 8 and up 75% of cases occur in healthy children and 25% associated with abnormalities of the development of the urinary tract and other malformations.

The majority of them are unilateral (95%), more frequent in the upper pole of the affected kidney. They tend to be encapsulated and vascularized, they do not usually exceed the abdominal midline towards their opposite side. They tend to metastasize to the lung.

It has been shown that 30% of patients studied with Nephroblastoma had a gene inactivated on the X Chromosome called WTX.

It responds very well to medical-surgical treatment. The survival rate is 90% after 5 years.

METHODOLOGY

A study of a pediatric patient with a right renal tumor lesion was performed, who underwent clinical-surgical treatment

with post-treatment follow-up.

The information obtained rests on the Word computer system and the image of those who carried out the study.

CLINICAL CASE PRESENTATION

3-year-old female patient.

As an important family history, it should be noted that the girl's mother was diagnosed, operated on and treated for a diagnosis of bilateral Wilms tumor in her childhood; free of disease and routine checks.

Parents refer clinical picture for approximately 30 days characterized by asthenia and constipation; Due to maternal experience and knowledge that it may be genetic, they go to the doctor who requested the Axial Computed Tomography of the Abdomen where there was evidence of a right renal tumor in the upper pole; deciding to go to a specialist.

Physical examination as positive shows a lesion in the right hemiabdomen that does not reach the abdominal midline. Staging examinations were carried out to decide on therapy, showing a tumor limited to the upper pole of the right kidney, with an intact renal capsule, classifying it as resectable in Stage I.

Nephroureterectomy + Appendectomy was performed; Uncomplicated surgical procedure with satisfactory post-

surgical recovery.

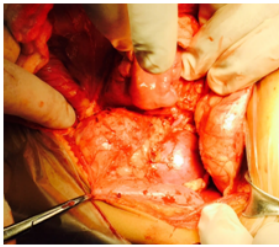


Image 1: Trans-surgical, exposure of tumor lesion



Image 2: Surgical Piece (Bluish region corresponding to tumor)

Histopathological result compatible with Nephroblastoma, well developed cellular components. Subsequently, the patient underwent chemotherapy treatment for 18 weeks.

At the moment we are 4 years after finishing treatment, patient is without evidence of recurrent disease, maintaining routine controls.

DISCUSSION

Second most frequent pediatric abdominal tumor. Obligatory to know extent of the disease for therapeutic planning. The removal of tumor and adjacent tissues is necessary and very important. To this, the administration of radiotherapy and adjuvant chemotherapy as treatment obtain survival rates of 80 - 90%.

CONCLUSION

Common pediatric abdominal tumor, the second after Adrenal Neuroblastoma. Timely detection, with correct staging and adequate treatment is the objective to achieve control of the disease.

Complete tumor removal with adjacent compromised tissue is the primary goal of treatment to improve prognosis, even achieving cure.

REFERENCES

1. Maiti S, Alam R, Amos CI, Huff V (2000). «Frequent association of beta-catenin and WT1 mutations in Wilms tumors». *Cancer Res* 60 (22): 6288-92. PMID 11103785.
2. Rivera M, Kim W, Wells J, Driscoll D, Brannigan B, Han M, Kim J, Feinberg A, Gerald W, Vargas S, Chin L, Iafrate A, Bell D, Haber D (2007). «An X chromosome gene, WTX, is commonly inactivated in Wilms tumor». *Science* 315(5812): 642-5. PMID 17204608. doi:10.1126/science.1137509.
3. Ruteshouser EC, Robinson SM, Huff V (junio de 2008). «Wilms tumor genetics: mutations in WT1, WTX, and CTNNB1 account for only about one-third of tumors». *Genes Chromosomes Cancer* 47 (6): 461-70. PMID 18311776. doi:10.1002/gcc.20553.
4. Richard E. Behrman, Robert M Kliegman, Hal B. Jenson. Nelson Tratado de Pediatría (en español). Publicado por Elsevier España, 2004; pág 1713. ISBN 8481747475
5. American Academy of Family Physicians (abril, 2001). Recognition of Common Childhood Malignancies (artículo completodisponible en inglés). *Am Fam Physician*; 61:2144-54. Último acceso 18 de febrero, 2009
6. M J Coppes, G J Liefers, P Paul, H Yeger, and B R Williams. Homozygous somatic Wt1 point mutations in sporadic unilateral Wilms tumor (artículo completodisponible en inglés). *Proc Natl Acad Sci U S A*. 1993 February 15; 90(4): 1416-1419. Último acceso 8 de abril, 2009. PMID: PMC45884
7. Paulino, Arnold C; Max J Coppes (diciembre de 2006). «Wilms tumor». *Pediatrics: General Medicine >Oncology* (en inglés). eMedicine.com. Consultado el 29 de enero de 2009.