

Nephroblastoma in Adults about a Clinical Case

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How to cite this paper: Cheickna, T., Amara, C., Amadou, M.O., Coulibaly, B.B., Daye, B.K., Hamidou, S., Gabriel, B.H.J.-B. and Lamine, D.M. (2024) Nephroblastoma in Adults about a Clinical Case. *Surgical Science*, 15, 81-88.

<https://doi.org/10.4236/ss.2024.152009>

Received: August 19, 2023

Accepted: February 26, 2024

Published: February 29, 2024

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Abstract

Nephroblastoma is the most common malignant renal tumor in children and is related to an abnormal proliferation of cells resembling those of the embryonic kidney (metanephroma), hence the terminology; embryonal tumor. These are tumors that remain and remain unstudied in Mali because they are common in adults in our context. Its annual incidence is estimated at approximately 1/10,000 births. Nephroblastoma is a rare or even exceptional tumor in adults. The clinical manifestation was a large swelling of the right hypochondrium; abdominal pain for a year; unquantified fever, hypertension, initial hematuria associated with burning during urination and anemia. The main clinical manifestation remained fever and abdominal pain. This renal tumor posed a diagnostic problem which was previously labeled as a mesenteric tumor in our general surgery department. The diagnosis was made by imaging: CT and magnetic resonance imaging. The treatment is multidisciplinary and combines chemotherapy, surgery with or without radiotherapy. The prognosis is poor due to late diagnosis and less effectiveness of chemotherapy compared to the child. Survival did not exceed a year and a half because the renal tumor in our patient was surgically overcome. We report a case of nephroblastoma in an 86-year-old patient with unfavorable histology (hematogenous metastases), operated on in the general surgery department and whose postoperative course was simple and who was referred to oncology for treatment.

Keywords

Nephroblastoma, Malignant Kidney Tumor, Treatment, Adult, Mali

1. Introduction

Nephroblastoma is the most common malignant renal tumor in children [1]. It

represents more than 90% of kidney tumors in children and approximately 5 to 14% of all pediatric and exceptional cancers in adults [2] [3] [4]. Kidney cancer is the third most common urological cancer after prostate and bladder cancer [3]. The annual incidence of nephroblastoma is estimated at approximately 1/10,000 births and it affects both boys and girls [1] [3].

Clinically an abdominal mass (unilateral in the majority of cases) is frequently present. Patients sometimes suffer from abdominal pain (around 10% of cases), hypertension, fever (20% of cases), hematuria and anemia. The disease is very rapidly progressive with regional dissemination in the retroperitoneal space, the lymph nodes, the vessels (renal vein and inferior vena cava), and in the peritoneal cavity in the event of tumor invasion and has a strong metastatic potential to the lungs and the liver [5] [6].

Nephroblastoma is sporadic, 10% of which are associated with congenital anomalies (aniridia, hemihypertrophy, urogenital anomalies) and are part of specific syndromes (Beckwith-Wiedemann, Denys-Drash, WAGR, Perlman syndromes). We find genetic, chromosomal anomalies: 11p13 (containing the WT1 gene), 11p15.5 (containing the H19 gene), 16q, 1p and 17p. Familial forms are exceptional (1% of cases) and are transmitted autosomal dominantly [2] [7].

Diagnosis is based on imaging, CT or magnetic resonance imaging. The dosage of urinary catecholamine metabolites is normal. It makes the differential diagnosis with mesoblastic nephroma (in infants), clear cell sarcoma, neuroblastoma (invasion of the kidney by contiguity), rhabdoid and metanephric stromal tumors [2] [3] [5] [8].

The treatment is multidisciplinary, combining chemotherapy, surgery with or without radiotherapy. Microscopic examination makes it possible to confirm the nephroblastoma and to specify the stage of the tumor determining the choice of post-operative chemotherapy. Radiotherapy is reserved for more extensive forms or with more unfavorable histology [4] [5] [8] [9].

The prognosis is favorable early diagnosis with survival greater than 90% of cases [10].

In our context, the scarcity of diagnostic means, the low specificity of symptoms, the low level of education of the population and the lack of screening programs mean that these cancers are often discovered at advanced stages. The aim of our work is to highlight the benefit of early diagnosis, differential diagnosis with typical Wilms tumor, and the benefit of initial surgical treatment due to chemoresistance of the tumor.

The diagnosis, often histological, is generally made at a later clinical stage than in children, the prognosis at this stage in children being similarly more pejorative in adults.

The aim of our work was to highlight the interest of imagery; the differential diagnosis with mesenteric tumor and the benefit of initial surgical treatment due to the chemoresistance of the tumor.

We report a case of stage IV nephroblastoma (hematogenous metastases), with unfavorable histology in an 86-year-old patient operated on in our general

surgery department, previously diagnosed for mesenteric tumor and whose operative outcomes were simple and who was referred to oncology for chemotherapy.

2. Observation

D.F, 86 years old, female, housewife consulted for a large abdominal swelling, predominantly in the right hypochondrium associated with abdominal pain for more than a year, stabbing type, intermittent late post-prandial accompanied by nausea with fever not quantified, calmed by taking painkillers and a gastric bandage.

The urinary signs were dominated by burning during urination with cloudy urine, pollakiuria, and initial hematuria at the beginning occurring a few times. Eating habits and medical-surgical history are unremarkable apart from a multiparity (G9P9V4D5A0) and lives in a gold zone of uncontrolled use of gold processing chemicals in Kéniéba, Kayes in Mali.

His general condition is acceptable with blood pressure = 10/7 cm Hg, respiratory rate at 22 cycles/min, pulse at 80 beats per minute and temperature at 38.2°C. The conjunctivae are moderately colored and consciousness is good.

Clinical examination revealed a large abdominal tumor located in the right hypochondrium occupying the hypogastrum, hard and mobile in relation to the deep plane. The rest of the examination is normal, no ascites. With regard to the questioning and the clinical examination, we carried out additional examinations to establish our diagnosis because the symptomatological description by the patient was confusing with a probably mesenteric digestive tumor.

Abdominopelvic ultrasound showed a heterogeneous necrotic tissue mass on the right flank (digestive) measuring 216 × 162 × 148 mm with a malignant appearance associated with thrombosis of the inferior vena cava compatible with vascular metastasis, a large poorly differentiated echogenic right kidney with a cortical cyst of 33 millimeters.

The electrocardiogram revealed a regular sinus rhythm with an anterior hemiblock. Cardio-Doppler ultrasound showed concentric remodeling of a relaxation anomaly which did not contraindicate surgical intervention.

Abdominopelvic tomography revealed a necrotic mass in the right renal compartment related to renal cell carcinoma and invasion of the inferior vena cava associated with suspicious left lumbo-aortic lymphadenopathy.

Biology showed an ESR = 74 millimeters at the 1st hour (N<15 mm); Creatinemia = 1.3 mg/dl; Rh group = O+ (positive); TP/INR = 1.0; Fasting blood sugar = 0.85 g/dl; Thick film positive for 50 trophozoites/mm³ of blood; NFS: GR = 3.21 million/microliter; GB = 6.6 × 10³/microliter; Hematocrit = 24.3%; Hemoglobin = 7.4 g/dl; VGM = 76 μm³ TCMH = 23.2 μm³; Platelets = 517 × 10⁶/microliter; Lymphocytes = 26.8%; Monocytes = 0.5 and Eosinophils = 0.8 × 10⁹/l. The diagnosis of right nephroblastoma was made.

Under general anesthesia with orotracheal intubation we performed a median laparotomy above and below the umbilical (**Figures 1-3**), an enlarged neph-

rectomy (**Figure 4**) removing the ipsilateral metastases associated with an ablation of the right ureter and a biopsy of the operating room. The postoperative course was simple (**Figure 5**).

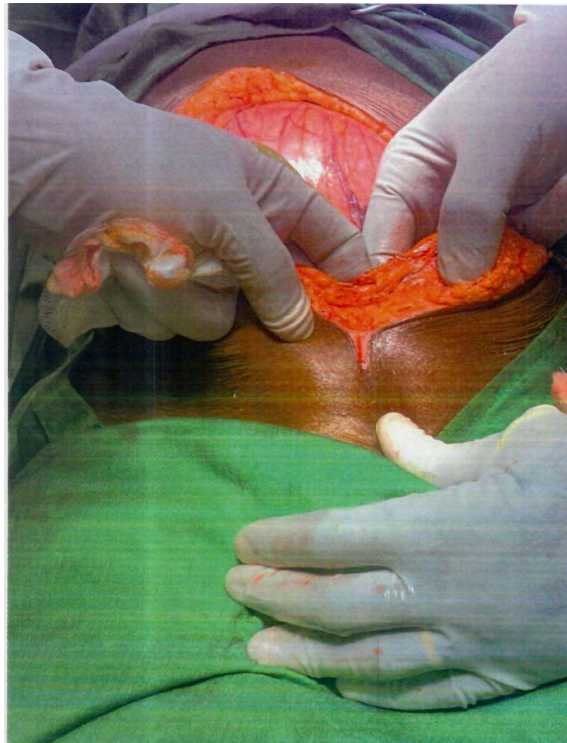


Figure 1. Enlargement of the laparotomy.



Figure 2. Enlargement of the laparotomy.



Figure 3. Tumor exploration.



Figure 4. Operating room.



Figure 5. Simple operating sequence.

The biopsy concluded that it was a papillary carcinoma of the right kidney grade 3 according to ISUP and presence of necrosis. The hilum and ureter were infiltrated by the tumor with the presence of vascular emboli and peri-renal fat, stage pT3aNxMx. She was referred to oncology for better care.

3. Discussion

Nephroblastoma, an embryonal tumor of the kidney, develops from the blastema of the metanephros normally under the induction of bud born on the metanephric duct differentiating into the tubule and glomerulus. Blastema proliferation leads to the formation of a tumor made up of young, undifferentiated cells when blastema differentiation (stopping around the 34th week and persisting at birth) does not occur [3] [9].

The incidences are relatively comparable, showing the rarity of this malignant kidney disease [5] [6] [11]. Currently there is no pediatric urology service in Mali. The studies carried out found an average age of 48.09 ± 10 years as in Morocco (52 years), Senegal (49 years), Nigeria (47.5 years) and the same in India (55 years) [5].

According to the literature, nephroblastoma represents 20% of abdominal tumors and shows a sex ratio favoring the male sex [2] [6] [12]. The etiologies are multiple and varied apart from congenital and genetic anomalies [11] [12] [13] [14]. Chemical substances are also implicated in the occurrence of cancers [7]. Our patient lived in a gold mining area with uncontrolled use of chemical

substances for gold processing in Kéniéba in the Kayes region of Mali and which flood the waterways, always criticized by the local population.

The clinical symptoms in adults are dominated by an abdominal mass, lower back pain and hematuria [3]. Non-urological signs may be encountered, namely high blood pressure, long-term fever or acute abdominal syndrome.

The biology is dominated by anemia, polycythemia, accelerated sedimentation rate and elevated creatinemia [6]. Other authors report an elevation of lactate dehydrogenase (LDH) in correlation with the size and stage of the nephroblastoma [8].

The diagnosis is based on imaging, namely ultrasound, uroscanner and/or computed tomography, confirmed by histology. Ultrasound makes it possible to specify the site, the echogenicity of the tumor, its local impact and an interest in the extension assessment. Doppler ultrasound can detect arterial and/or venous signs suggestive of tumor neovascularization [4] [9]. Ultrasound, computed tomography and magnetic resonance imaging have recently popularized the indications and management [2] [3] [4] [5] [8] [9].

Treatment is multidisciplinary based on surgery and chemotherapy. This chemotherapy can be pre [4] and post operative according to the authors [4] [6] [8] [10]. Radiotherapy is reserved for progressive stages III and IV of nephroblastoma [10]. We recommended surgery and after chemotherapy for our patient as with many other authors [2] [3] [5] [6] [8] [9].

The prognosis depends on the stage of the nephroblastoma [12] [13] [14] [15]. It is unfavorable in the majority of cases because it is discovered too late [2] [6] [8] [9] [10] [12] like the case of our patient.

4. Conclusion

Nephroblastoma or Wilms tumor in adults today constitutes a very rare kidney cancer in Mali, especially at this age (86 years) due to its rapid progression with a high metastatic potential. The first sign is hematuria, considered trivial and is attributed to urinary bilharzia. Abdominopelvic ultrasound, computed tomography and especially magnetic resonance imaging have recently popularized or improved diagnosis and operative indications. However, the prognosis is always reserved because it depends on the histology and the progressive stage of the kidney tumor.

Acknowledgments and Conflict of Interest

The authors thank the patient who kindly gave her informed consent and there is no conflict of interest in relation to this article.

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