

Bilateral Nephroblastoma: A Case Report On A 4-Year-Old Child, Imaging, Pediatrics, And Pediatric Surgery Department At The Panzi General Reference Hospital, Bukavu, South Kivu, Democratic Republic Of Congo, Patient Hospitalized January 2022

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Abstract

Bilateral nephroblastoma accounts for 4-8% of all Wilms tumors (WT). Nephroblastomatosis (NB) is characterized by the presence of numerous or diffuse nephrogenic rests (NRs). These lesions are always present in bilateral nephroblastoma and can be better recognized using computed tomography (CT). Treatment is a comprehensive strategy that includes chemotherapy, surgery, and radiotherapy. Bilateral nephroblastoma accompanied with NB is uncommon, leading to inconsistent treatment options and a lack of standard methods and facilities in our region.

A 4-year-old boy died from bilateral nephroblastoma due to inadequate treatment and facilities. A new protocol adaptor tailored to the reality of developing countries would be able to address this issue.

Keywords: *Bilateral nephroblastoma, 4 year old boy, Nephroblastomatosis*

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I. Introduction

Nephroblastoma is an embryonic tumour that develops in early life. It works particularly well in grownups. Nephroblastomas account for about 95% of all pediatric kidney tumors. Bilateral localization, whether immediate or secondary, is uncommon, accounting for 4-8% of all WTs. This form appears in younger children and is commonly connected with NB.(1,2,3)

The presence of NRs is the defining feature of nephroblastomatosis. In unilateral WTs, NRs are typically only discernible through histology, whereas in bilateral WTs, they can be observed on imaging. Abdominal ultrasound and chest X-rays are employed as the first-line diagnostic tools. To confirm the an ultrasound results, an additional histology is conducted.(6,8,9)

We report the case of a four-year-old boy admitted for bilateral nephroblastoma with NB. The purpose of this study is to demonstrate the difficulty of interpreting imaging data that lead to diagnosis, as well as the lack of diagnostic and therapeutic options , modalities and methods in low-income countries.

II. Case Report:

A four-year-old boy was admitted with an abdominal mass. The clinical examination revealed a hard mass in the left hypochondrium that spread to the left flank and right hypochondrium, as well as stomach pain, cough, fever, hematuria, high blood pressure, loss of appetite, nausea, and vomiting. Biologically, the patient showed increased lactate dehydrogenase (600U/L), CPR (20mg/dl), and hemoglobin (18g/dL).

The abdominal ultrasound revealed a hypochoic and heterogeneous bilateral renal mass, as well as a hypochoic corticomedullary nodule related to NB lesions measuring 15 cm on the left and 5 cm on the right. The abdominal CT scan and IRM were not available. A chest x-ray revealed cannonball metastasis, which is defined as numerous big, well-circumscribed (balloons release images of both lungs) lung metastases.

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Following a multidisciplinary concentration meeting, the boy received preoperative and palliative care. After two weeks of observation, the patient was operated on; a preoperative procedure and anaesthetic protocol were developed; surgical exploratory laparotomy exploration revealed bilateral nephroblastoma with NB.

The left tumor was removed by subtotal nephrectomy (6 cm), and the right upper pole tumor was resected with an ultrasonic knife. Both samples were sent to anatomopathology and histology for deep assessment, and the histological findings were bilateral NB with a multi-centric WT of favorable histology on the left side and peripheral on the right side.

III. Discussion

Kidney tumors make about 5-8% of juvenile malignancies and are more prevalent in children under 6 years of age(2). Approximately 95% of these tumors are nephroblastomas. Bilateral nephroblastoma accounts for 4–8% of cases(4,7,9).

The presence of many or diffuse NRs denotes nephroblastomatosis. In unilateral nephroblastoma, they occur in 12-17% of nephrectomy pieces, but in bilateral nephroblastoma, they can occur in up to 100% of cases. This shows that NRs may be a precursor lesion of nephroblastoma, as they have a similar physiopathogenic phenomena(4,5,8).

Two types of NR are recognized: intralobar (ILNR) and perilobar (PLNR). Perilobar nephrogenic rest occurs in 1% of pediatric autopsies, while intralobar form is observed in 0.1%. In unilateral WT, NRs are typically only discernible through histology, while in bilateral WT, they can be observed on imaging(2,7).

The presence of NRs after nephrectomy for nephroblastoma increases the chance of contralateral tumor. The danger is higher for children under one year old(6).

Nephroblastoma can be suspected clinically and radiologically, but histology is required for confirmation(3).

Nephroblastoma might present as stomach pain or a palpable lump. Rarely, patients may experience hematuria, fever, or elevated blood pressure(5).

Abdominal ultrasounds and chest X-rays are employed as the primary diagnostic tools. A CT scan or an MRI is performed a second time to validate the ultrasound results, but in our instance, all of the specified exams were unavailable and the family did not have the necessary financial means. An abdominal and pelvic CT scan is performed before and after contrast administration during the portal phase. If suspicious lesions appear on the chest X-ray, a chest CT scan is performed during the injection acquisition(5,6).

Imaging is used to confirm the diagnosis of nephroblastoma and identify any related abnormalities, such as "horseshoe" kidneys or other urogenital anomalies, which can impact treatment options.

Imaging findings for nephroblastoma vary depending on the tumor's components (necrosis, hemorrhage, calcification, and fat). Characteristic indicators include a well-defined solid mass with a pseudocapsule, resembling a "claw" of renal parenchyma around the tumor(6).

In pediatrics, ultrasound (US) is typically used as the initial line of inquiry for abdominal masses. Nephroblastomas present as solid, heterogeneous masses with calcification, but are rarely cystic on ultrasound.(5).

Ultrasound imaging of nephroblastomatosis might be hypoechoic, hyperechoic, isoechoic, anechoic, nodular, or diffuse. Computed tomography is more effective than ultrasound in detecting nodular or diffuse isodense lesions that do not respond to contrast injection. MRI reveals iso-intense NB lesions that become hypo-intense in T1 after gadolinium administration(6).

Nephroblastoma is characterized by three phases in histology : blastema, epithelial differentiation, and stromal differentiation, in varying proportions.

Treatment consists of a comprehensive therapeutic approach that includes chemotherapy, surgery, and radiotherapy. The rarity of bilateral nephroblastoma coupled with NB explains the variety of therapeutic approaches, as well as the lack of codified procedures and a competent technical platform, which led to symptomatic and palliative treatment following diagnosis in this instance. Management must be multidisciplinary and respond to two opposing imperatives: be as curative as possible while preserving the maximum renal parenchyma to avoid progression to renal failure. Bilateral nephroblastoma is often treated with preoperative chemotherapy, surgery, and postoperative chemotherapy, which may also include radiotherapy(1,9).

IV. Conclusion

Bilateral nephroblastoma is characterized by nephroblastomatosis lesions. Although CT scans are more effective, abdominal ultrasounds can also reveal these abnormalities, as seen in our instance. Bilateral NB management requires a multidisciplinary consultative meeting to make the final therapy decision.

Faced with a shortage of human resources and equipment in underdeveloped nations, such as ours, we are limited, even in terms of diagnostic tools. We can only limit ourselves to the resources available to us. To avoid such drifting development, we recommend establishing a prenatal and perinatal diagnostic platform, as well as providing oncology treatment centers accessible to all, within the limits of available resources, with multi-partner public-private support, and accelerated training in diagnosis and general care in pediatric oncology for all healthcare personnel, according to their functions and responsibilities. Because we had a treatment alternative but limited the technical and monetary means, we were willing to provide palliative care to the child, which was tremendously destructive to us. As long as we can detect early and have therapy alternatives accessible, the evolution will be favorable. It is consequently critical that all stakeholders—governments, NGOs, and healthcare professionals—are aware of and accountable for tackling this pandemic. The most responsible answer is to implement a well-established multidisciplinary healthcare routine that is appropriate for developing countries and accessible to everybody.

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