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Surgical Treatment of Nephroblastoma in a 4-Year Male Child: A Case Report and Literature Review

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Abstract

Herein we report a patient with nephroblastoma which was successfully removed at the Sunyani Teaching Hospital CJ Oppong theatre in Ghana in sub-Saharan Africa by extended below umbilical incision. Our patient had a family history of Wilms tumour predisposing him to the disease. His main symptoms were haematuria and abdominal mass which was noticed later. Examination and investigation were suggestive of a late-stage unilateral Nephroblastoma resulting in the need for nephrectomy. To reduce the need for such radical surgeries among children at an early stage, there is the need for early screening of children for Wilms tumours especially, those with family predisposition as in our case study. The case report presented here constitutes a rare case of nephroblastoma in the literature.

Keywords

Nephroblastoma, Wilms Tumour, Nephrectomy, Screening

1. Introduction

Wilms' tumour is the common renal malignancy in children less than five years old and it accounts for approximately 95 percent of all cases diagnosed before the age of four with the peak incidence in the age range of 2 - 5 years [1] [2]. Primarily, Wilms' tumour is a sporadic disease and only 1% to 2% of patients with Wilms tumour have a relative with the disease [3] [4]. In approximately 10 percent of cases, Wilms tumour occurs as a part of a multiple malformation syndrome, including WAGR, Denys-Drash, and Beckwith-Wiedemann syndrome

(BWS) [2] [5] [6] [7]. Epidemiologically, Wilms can present as a solitary tumour, bilateral kidney involvement or a multifocal locus in a single kidney with the distribution of the majority being as a solitary tumour and 5 - 7 percent, 10 percent respectively for the other forms. Ethnically Wilms tumour is more common among African Americans and less among Asians. Wilms tumour appears to be caused by abnormal renal development, resulting in proliferation of the metanephric blastema without normal tubular and glomerular differentiation [8] [9]. Most children with Wilms tumour present with an abdominal mass or swelling, without other signs or symptoms. Other symptoms can include abdominal pain (30 to 40 percent of patients), haematuria (12 to 25 percent), fever, and hypertension (25 percent) [10]. A subset of patients with subcapsular haemorrhage can present with rapid abdominal enlargement, anaemia, hypertension, and, sometimes, fever.

Screening for Wilms tumour can be done for high-risk patient specifically those with family pre-deletion and children with congenital disease which mostly have Wilms tumour associated with example WAGR syndrome when screening done by abdominal ultrasound can help pick it up.

The diagnostic and evaluation of Wilms tumour needs a clinical approach including taking a detailed history, examination and investigations. Wilms tumour is confirmed by histopathology but initial evaluation includes abdominal ultrasound CT scan chest x-ray which will help stage the tumour, identify metastasis, complications of the tumour so as to help prepare for surgery and plan the treatment modality.

The staging and management of Wilms tumour are based on two criteria: the Children's Oncology Group (COG) and the International Society of Paediatric Oncology (SIOP).

These criteria are based on the relation of surgery and chemotherapy. The COG surgery is done before chemotherapy and tumour staged while in the SIOP chemotherapy is done and surgery afterwards and also based on whether there is unilateral renal involvement or bilateral renal involvement [2] [10]. The management consists of surgery, chemotherapy and radiation. The options chosen are mainly dependent on the modality of SIOP or COG and the stage of the disease. This staging shows the intensity of chemotherapy if warranted and the need for radiotherapy. Overall, 5-year survival between SIOP and COG is not very wide.

Below is a case of a 4-year-old boy with Wilms' tumour with mild anemia.

2. Case Presentation

A 4-year-old boy was the only child of both parents. The child lived with his father. The child was never admitted into the neonatal intensive care unit. There was no history of neonatal sepsis. The developmental milestones were appropriate for his age. The child received full vaccines up until the current hospitalization. The child was in his usual state until six months ago, when was noticed to have blood in his urine, and was sent to a peripheral health facility where he was

given some medications and it stopped. Three months after, the child was noticed to have an abdominal mass at the left flank and was sent to a peripheral health facility where upon examinations and investigation, client was referred to our facility (Sunyani Teaching Hospital).

On direct questions, there was no hematuria, weight loss, or abdominal pain.

3. Physical Examination

The child looked clinically stable, well-nourished, afebrile not pale and hydration was fair. The child's temperature was 37˚C, pulse 99 bmp, respiratory rate 20 cpm, SPO_2 99%. Heart sound S1 and S2 present, no murmur head, air entry adequate, vesicular breath sound; no added sound heard. Gastrointestinal tract was normal. The child was feed adequately with normal bowel habits.

4. Clinical Examination

At the time of examination, the abdomen was soft and not tender. At the left-side of the abdominal wall was a palpable mass with smooth surface non-tender, fair not fluctuant of size 10 by 8 cm.

5. Investigations

Abdominal CT report

Examination: Contrast enhanced CT scan of the Abdomen on 19/04/2023

Report:

Serial axial CT scan images of abdomen shows a heterogeneously enhancing 11.7 × 7.9 × 9.6 cm mass arising from the inferior pole of the left kidney with calcifications and evidence of stretched renal parenchyma along its periphery and normal upper pole renal tissue. The mass is noted to cross the midline over the abdominal aorta. There is no evidence of encasement of infiltration into vessels. The right kidney shows normal size and position. The renal parenchyma shows normal width and structure, no evidence of a focal lesion or calculi, normal liver architecture and no hepatomegaly. The intrahepatic and extra-hepatic ducts and gallbladder are unremarkable. The pancreas is with normal size position and internal structures with smooth lobulated margins. Pancreatic duct is unobstructed. The adrenal glands are unremarkable. Major blood vessels appear normal and there is no evidence of lymphadenopathy. Imaged portions of lungs and soft tissues are normal.

Impression: Findings consistent with left Nephroblastoma as described above.

CT scan films of left nephroblastoma as **Figure 1** below.

Laboratory Biochemistry test with reference ranges is shown in "**Table 1**" below. Parasitology (Urine R/E) and Haematology results with reference ranges are also shown in "**Table 2**" below.

6. Surgery

Child was operated on 16th May 2023. Surgery was done under general anesthesia,

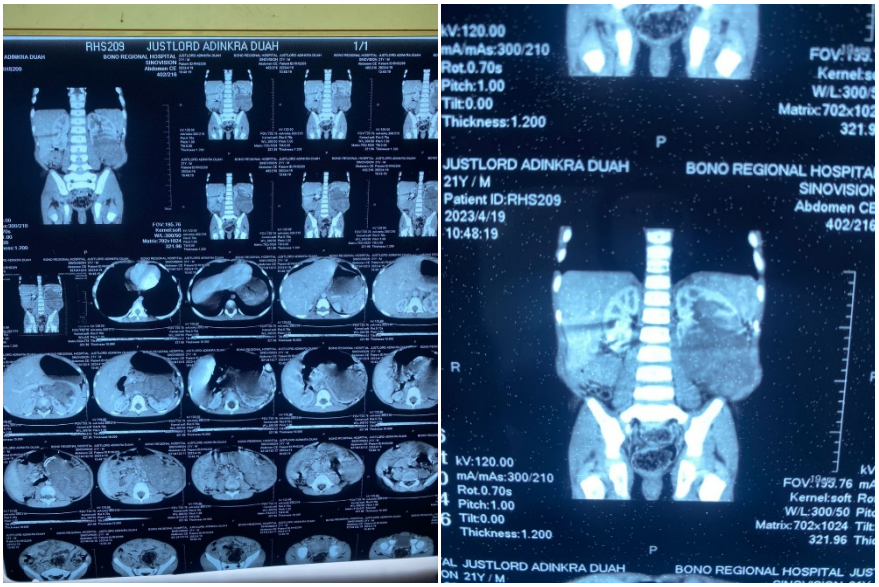

Figure 1. CT scan of kidneys.

Table 1. Chemistry test.

RFT				
Test Name	**Results**	**Flag**	**Reference range**	**units**
SODIUM	135.9		135 - 145	mmol/l
POTASSIUM	4.66		3.4 - 5.5	mmol/l
CHLORIDE	106.5		95 - 109	mmol/l
UREA	4.4		1.7 - 8.3	mmol/l
CREATININE	38.8		Female: 53 - 108 Male: 62 - 115	mmol/l

Table 2. Parasitology (Urine R/E).

Macroscopy	Appearance: Clear Colour: Straw
Chemistry	Leucocytes: Trace; Nitrite: Neg; Urobilinogen: Normal; Protein: Neg; PH: 6.0; Blood: Neg; SP Gravity: 1.0λg; ketones: Trace; Bilirubin: Neg Glucose:
Microscopy	Pus Cells per high power field: 20; RBCs " " " " ": 0 Epith Cells " " " " ": 4 S. haematobium ova: T. Vaginalis: Candida: …… Not seen…… Cast: ……
Haematology	HB 9.2 g/dl. WBC 8.75 × 10^9/L, MCV 75.2 fl, platelet count 252 × 10^9/L

child was well draped; a midline skin incision done, subcutaneous tissues and fascia divided, abdomen was entered, left radical nephrectomy done, haemosta-

sis secured, there was minimal blood loss about 100 mls. Intra operative was normal looking intra-abdominal structure with exception of the left nephroblastoma, no metastasis or infiltration of vessels seen. Surgery was successful. The child was sent to recovery ward after surgery and was later transferred to the paediatric surgical ward after the recovery from the anesthesia. The child was given IV Ceftriaxone 500 mg bd, intravenous fluid which was started pre operatively and continued, Paracetamol Supp. 250 mg 6 hourly for 48 hours. The performance of surgery is shown in Figure 2 and Figure 3.

Gross dissection of nephroblastoma is shown in Figure 4 and Figure 5 below.

The nephroblastoma has taken the whole kidney with no parenchyma tissue left as shown the above.

Figure 2. Surgery in progress.

Figure 3. Surgical team.

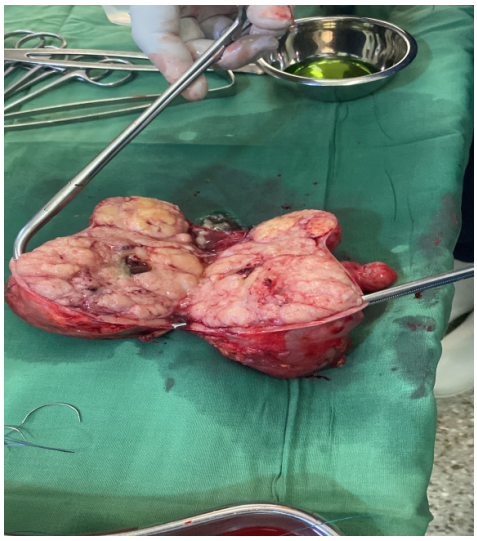

Figure 4. Cross section of dissected kidney.

Figure 5. Left kidney.

7. Histopathology Report on Nephroblastoma of Left Kidney

Stage 1 (9 × 8 cm tumor centrally located which grossly involves the pelvis and mostly upper and pushing down on the lower pole. evident infiltration in renal capsule).

Histopathological Diagnosis

Nephrectomy with proximal ureter triphasic nephroblastoma without anaplasia

Other features

Tumour size: 9 cm

Necrosis in tumour tissue: not present

Multifocality: not present

Infiltration into renal capsule: evident

Resection margins in ureter: free
Renal capsule appears intact on microscopy
Number of limp nodes examined: 0
Stage (NWTS): 1
pTNM classification: pT2NxMx

8. Post Operative Reviews

Post Op Day 1

Child had no complaints; vitals were normal and wound dressing was not soaked. Plan was to continue current treatment and start sips.

Post op Day 2

Child was doing well and ambulating. Syr. Amoxiclav 228 mg/5mls, 5 mls BD was added to medication and to continue sup paracetamol.

Post op Day 3

Wound dressing was opened; surgical site was clean and dry. Wound dressing was done and child was discharged on Syr paracetamol 10 mls tds and Syr Amoxiclav 228 mg/5mls 5 mls bd for seven days and scheduled for weekly OPD review. Child was later referred to Oncology Directorate at Komfo Anokye Teaching Hospital in Kumasi for Chemotherapy. Patient is currently doing very well and parent very happy.

9. Discussion

Wilm's tumour can grow for a long time without any characteristic symptoms, causing only fever, abdominal pain, nausea, or vomiting, which is the reason why it is often discovered accidentally [11]. In our case, the child had haematuria for which she was sent to hospital and treated and haematuria stopped. The most typical recognition of Wilms' tumour occurs when swelling starts in the abdomen, but by this point they have often grown quite large [12]. In our case, the child was noticed to have an abdominal mass at the left flank three months later after first haematuria.

It can be found earlier if the child gets ultrasound of the abdomen and confirmation by CT scan [13]. The full medical history should be taken to find out all symptoms that may relate and how long they have existed. To help with diagnosis of a tumour, ultrasound, imaging tests (x-rays, magnetic fields, or radioactive substances) are needed for differentiate diagnosis. These tests will help to define the location, type, and size of the tumour [13]. This imaging study helps to know the extent of the disease and also to plan or decide the treatment modality. In our case, CT scan was done, and there was no evidence of metastasis. Other test should be investigated such as haematological tests which in certain cases, child maybe will have different anaemia which might warrant haemotransfusion based on the severity and clinical presentation, but in this case the child had a mild microcytic anaemia, estimated blood loss was minimal during surgery so child was not haemotransfused. Hypertension was rare in children, if

the child gets high blood pressure, it could be another sign of a kidney tumour. Monitoring the child during the hospitalization is needed [14]. Blood and urine samples might also be collected for testing.

There is a familiar predation for Wilms's tumour even though this is rare, for those with a family history of Wilms' tumour or congenital abnormality there is a high risk of getting it. In this case, the child had no family history of congenital abnormality but a cousin from maternal side had similar condition and was operated on account of nephroblastoma. When the child is diagnosed as Wilms' tumour, tests can be done on parents [15]. Screening for Wilms' tumour is very important for children because it relates to child survival. Therefore, screening for Wilms' tumour is very important for the child who has birth defects, or a family history. A very simple and cheap way is for physical examinations to be done by a specialist; ultrasound on a regular basis is recommended; it could be done on a 3 - 4 months interval until the child reaches age of 7 years old [16].

For the management in this case COG guidelines were used where in our facility after initial assessment and the appropriate imaging studies done child was booked for elective nephrectomy which was done by the urology team. Intraoperatively, no metastasis was seen and histopathology stage malignancy as stage I with no anaplasia seen hence child was referred for chemotherapy with very positive results.

This decision was of benefit to our team based in sub-Saharan Africa where the management modality is based on case basis inasmuch that the surgeon chooses the best option for each case management hence able to benefit for both guidelines as can be seen above in the case.

10. Conclusion and Recommendation

The case was a 4-year-old boy with a family history of Wilms tumour, who presented with haematuria and abdominal mass which was noticed late, and examination and investigation were suggestive of a late stage unilateral Nephroblastoma resulting in the need for total nephrectomy. In order to reduce the need for such radical surgeries among children at an early stage, there is the need for early screening of children for Wilms tumour, especially those with family predisposition as in our case of study. We also recommend case specific management approach for each case of Wilms tumour.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

Consent

Consent for publication sought from patient and parent.

References

[1] Luu, D.T., *et al.* (2021) Wilms' Tumor in Horseshoe Kidney. *Case Reports in Neph-*

rology and Dialysis, **11**, 124-128. https://doi.org/10.1159/000514774

[2] Pater, L., *et al.* (2021) Wilms Tumor. *Pediatric Blood & Cancer*, **68**, e28257. https://doi.org/10.1002/pbc.28257

[3] Huff, V. (2011) Wilms' Tumours: About Tumour Suppressor Genes, an Oncogene and a Chameleon Gene. *Nature Reviews Cancer*, **11**, 111-121. https://doi.org/10.1038/nrc3002

[4] Tripathi, S., *et al.* (2021) Wilms' Tumor in Adults—Conventional and Unconventional Presentations of a Rare Entity with a Review of Literature. *Journal of Kidney Cancer and VHL*, **8**, 40-48. https://doi.org/10.15586/jkcvhl.v8i2.186

[5] Hol, J.A., *et al.* (2021) Clinical Characteristics and Outcomes of Children with WAGR Syndrome and Wilms Tumor and/or Nephroblastomatosis: The 30-Year SIOP-RTSG Experience. *Cancer*, **127**, 628-638. https://doi.org/10.1002/cncr.33304

[6] Mussa, A., *et al.* (2019) The Effectiveness of Wilms Tumor Screening in Beckwith-Wiedemann Spectrum. *Journal of Cancer Research and Clinical Oncology*, **145**, 3115-3123. https://doi.org/10.1007/s00432-019-03038-3

[7] Nelson, M.V., *et al.* (2021) New Approaches to Risk Stratification for Wilms Tumour. *Paediatrics*, **33**, 40-48. https://doi.org/10.1097/MOP.0000000000000988

[8] Spreafico, F., *et al.* (2021) Wilms Tumour. *Nature Reviews Disease Primers*, **7**, 75. https://doi.org/10.1038/s41572-021-00308-8

[9] Steur, A., *et al.* (2022) Characteristics and Outcome of Children with Wilms Tumor Requiring Intensive Care Admission in First Line Therapy. *Cancers*, **14**, 943. https://doi.org/10.3390/cancers14040943

[10] Groenendijk, A., *et al.* (2021) Prognostic Factors for Wilms Tumor Recurrence: A Review of the Literature. *Cancers*, **13**, 3142. https://doi.org/10.3390/cancers13133142

[11] Szymanska, A., *et al.* (2016) Wilms' Tumor with Intra-Atrial Extension: Treatment and Management. *World Journal for Pediatric and Congenital Heart Surgery*, **7**, 116-119. https://doi.org/10.1177/2150135115583728

[12] Hong, B. and Dong, R. (2021) Research Advances in the Targeted Therapy and Immunotherapy of Wilms Tumor: A Narrative Review. *Translational Cancer Research*, **10**, 1559-1567. https://doi.org/10.21037/tcr-20-3302

[13] Bălănescu, R.N., *et al.* (2022) Role of Ultrasonography in the Diagnosis of Wilms' Tumour. *Children*, **9**, 1252. https://doi.org/10.3390/children9081252

[14] Cunningham, M.E., *et al.* (2020) Global Disparities in Wilms Tumor. *Journal of Surgical Research*, **247**, 34-51. https://doi.org/10.1016/j.jss.2019.10.044

[15] Rutgers, J.J., *et al.* (2021) Interobserver Variability between Experienced and Inexperienced Observers in the Histopathological Analysis of Wilms Tumors: A Pilot Study for Future Algorithmic Approach. *Diagnostic Pathology*, **16**, Article Number: 77. https://doi.org/10.1186/s13000-021-01136-w

[16] Balis, F., *et al.* (2021) Wilms Tumor (Nephroblastoma), Version 2.2021, NCCN Clinical Practice Guidelines in Oncology. *Journal of the National Comprehensive Cancer Network*, **19**, 945-977. https://doi.org/10.6004/jnccn.2021.0037