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### Left Atrial Metastasis of Wilms Tumour in Abeokuta, Nigeria: A Case Report

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#### Abstract

**Background:** Wilms tumor (WT) is the most common renal malignancy, and also the second most common intra-abdominal cancer of childhood globally. It typically presents as an abdominal mass. In low-and-middle-income countries, however, patients tend to present with advanced disease which has extended beyond the renal bed. Evidence of metastasis has been reported in about 12% of WT cases, with the lungs being the most common site in more than 80% of advanced cases. Less frequently disease may spread to the liver, bones, brain, and rarely the heart.

**Objective:** To highlight left atrial metastasis as a rare complication of WT, as well as, emphasize the importance of echocardiography as a component of routine evaluation of suspected cases of WT.

**Methods:** We present a 5-year-old female who presented with a three-year history of left-sided abdominal swelling, one-year history of weight loss and recurrent abdominal pain, and three-week history of cough.

**Results:** Abdominopelvic CT scan and ultrasound were both in keeping with left nephroblastoma. Echocardiogram

revealed a left intra-atrial mass measuring 14mm X 17mm.

#### Treatment/Outcome

She was diagnosed with Stage IV nephroblastoma, commenced on neo-adjuvant chemotherapy. A repeat echocardiography was planned after the completion of the neo-adjuvant chemotherapy schedule. She however died during the 4th week of treatment.

#### Discussion

Wilms tumour has been reported to invade vascular structures with consequent extension to the right atrium and ventricle. Left atrial involvement in WT is a rare but documented complication of the disease, and could be an asymptomatic, incidental finding as it occurred in the index case.

#### Conclusion and Recommendations

Wilms tumour patients should have echocardiography done as part of their routine evaluation to provide baseline information about cardiac function prior to the commencement of potentially cardiotoxic medications, and also to screen for intra-cardiac extension of the disease.

**Keywords:** Nephroblastoma, Atrial Mass, Echocardiography, Case Report

#### Introduction

Wilms tumour (WT) is the most prevalent malignant renal tumor, constituting >90% of all renal cancers; it is also the second most common intra-abdominal cancer of childhood globally [1, 2]. In sub-Saharan Africa, WT is reported to be the most common solid tumor among children [3]. The typical clinical presentation of WT is an abdominal mass, with or without constitutional symptoms [4, 5]. In low and middle countries (LMICs) like Nigeria, however, patients typically present late, by which time the disease has extended beyond the renal bed [3, 4, 6]. Evidence of metastasis has been reported outside the abdominopelvic region in about 12% of WT cases, with the mode of spread being by direct extension, lymphatic or haematogenous spread [7] the lungs are the primary site for metastasis in more than 80%, with less frequent metastases to the liver, bones, and brain [7] and rarely the heart [8].

Intra-cardiac metastasis of WT, though rare, has been documented in literature [8, 9]. In a case series by Mapelli *et al.* [9], among 16 histologically confirmed WT cases, the prevalence of intra-cardiac extension was as high as 19%. The tumour has been reported to spread to the inferior vena cava (IVC) in about 4-10% of cases, the right atrium in approximately 1%, 9,10, and very rarely to the left atrium [11]. Patients with metastasis to the heart may be asymptomatic and identified during routine

echocardiography. Others may present with abnormal cardiac findings such as dynamic tricuspid valve obstruction, or cardiogenic shock [5, 6, 8, 9].

Wilms tumour is highly chemo-sensitive and curable, with 5-year overall survival (OS) and event-free survival (EFS) in developed countries approaching 90% with the current multi-modal treatment strategies. However, in most LMICs OS and EFS are less than 50% [4, 12, 13]. The relatively poorer outcomes in LMICs, are attributable to advanced disease stage at presentation as a result of poverty or poor health-seeking habits, lack of access to healthcare services, inadequate supportive care, and abandonment of treatment [3, 4, 12-15]. Globally, metastatic WT has a poorer prognosis compared to localized disease [5].

We present a five-year-old female with left atrial metastasis of WT to highlight this rare complication of the disease, as well as emphasize the importance of echocardiography as a component of routine evaluation of suspected cases of WT.

### Case Summary

AI was a 5-year-old female who presented to our centre with a three-year history of left-sided abdominal swelling, one-year history of weight loss and recurrent abdominal pain, and a three-week history of cough.

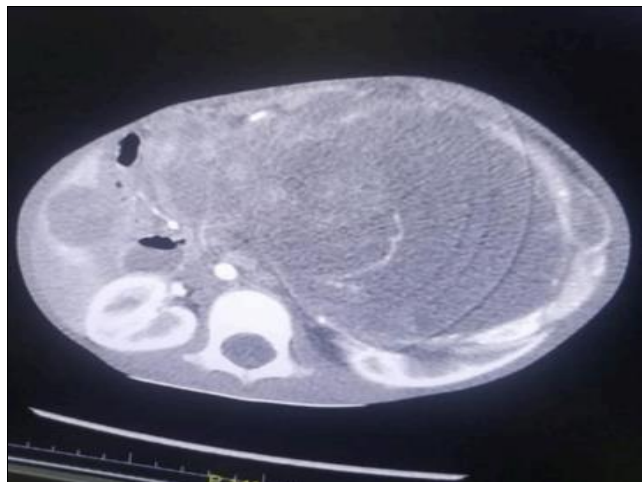
The abdominal swelling was insidious in onset, initially limited to the left flank but progressively increased in size to involve the whole abdomen. Two years later, she developed recurrent generalized abdominal pain, occasionally severe enough to disturb her normal activities; there were no known aggravating factors, but it was temporarily relieved with the use of analgesics. There was associated progressive weight loss, evidenced by the loosening of previously well-fitted clothes despite a good appetite. The cough was non-paroxysmal, non-barky with no post-tussive vomiting. She had been previously admitted at a private hospital a year before presentation where an abdominal ultrasound revealed an abdominal mass. She was subsequently referred to a tertiary facility, but her caregivers did not honour the referral as a result of financial constraints.

At presentation, she was conscious, cachectic, pale, not cyanosed, there was no pedal edema, but she had generalized lymphadenopathy. She weighed 13kg, while her weight for height was less than 3-Z-score. She had an unevenly distended abdomen with a prominent left flank swelling. There was a palpable left-sided mass occupying the left lumbar region and extending to the periumbilical area measuring about 17cm x 25cm, it was firm, non-tender, and not attached to the overlying skin. Her pulse rate and blood pressure were normal for age and height centiles, heart sounds were normal with no murmur.

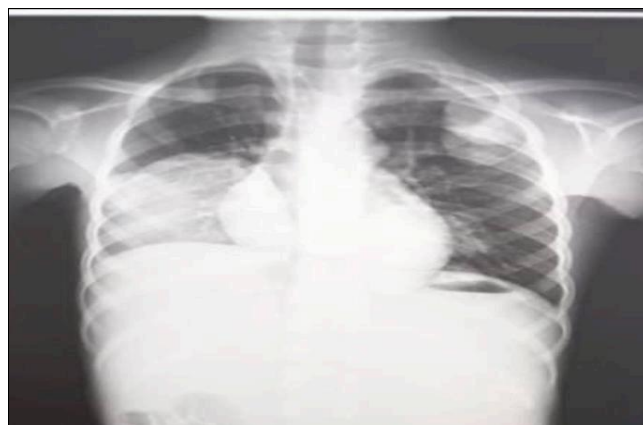
A presumptive diagnosis of WT (advanced disease) was made. She was admitted into the paediatric ward and commenced supportive treatment pending the outcome of her investigation results.

**Investigation results:** Abdominopelvic CT scan (Fig 1) and ultrasound were both in keeping with left nephroblastoma. Her chest radiograph revealed opacities suggestive of pulmonary metastasis. (Fig 2). Her initial haematology, hepatic, and renal function test results were essentially normal. An echocardiography was also requested as part of the routine evaluation, as per unit protocol, prior to commencement of neoadjuvant chemotherapy. Her echocardiogram (Fig 3) revealed a left intra-atrial mass measuring (14mm X 17mm) minimally attached to the

septal wall of the left atrium. The mass was uniformly echogenic with a smooth, rounded edge. There were no shunts, chamber dilatations, or hypertrophy. She however had a regurgitant tricuspid valve, though there was good ventricular function on both sides.



**Fig 1:** cT scan of the abdomen showing large left-sided intra-abdominal mass extending beyond the midline



**Fig 2:** Chest radiograph showing a homogeneous opacity involving the right middle and lower lung zones



**Fig 3:** 2D echocardiogram showing a left atrial mass measuring 14mm x 17mm

**Treatment and outcome:** She was diagnosed with Stage IV nephroblastoma, commenced on neoadjuvant chemotherapy, and scheduled to have a nephrectomy as per SIOP protocol<sup>[16]</sup>. A repeat echocardiography was planned after the completion of the neoadjuvant chemotherapy schedule. A multi-disciplinary team that included paediatric oncologists and cardiologists, cardio-thoracic surgeons, paediatric surgeons, and a palliative care unit was involved in her care. Other supportive care such as analgesia, fluid therapy, allopurinol, and blood transfusion were also instituted as appropriate. She however died during the 4th week of neoadjuvant chemotherapy.

### Discussion

Wilms tumour has been reported to invade vascular structures such as the hepatic veins and IVC, with consequent extension to the right atrium and ventricle<sup>[10, 17-19]</sup>. Very rarely, the left side of the heart may be involved<sup>[11]</sup>. Intra-cardiac extension of WT is often diagnosed radiologically following clinical suspicion, while some are discovered incidentally during routine work-ups,<sup>[17]</sup> as it occurred in the index patient.

Patients with intra-cardiac disease are usually asymptomatic, but they are at great risk of distant thromboembolism and death, hence, it should be considered in all children with WT<sup>[18, 19]</sup>. No relationship has been established between the size of WT and the presence of an intra-cardiac mass. Screening is therefore important for all categories of patients to limit disease-related morbidities and optimize care<sup>[19]</sup>.

Our literature search revealed that only a few cases of intra-cardiac extension of WT have been reported in sub-Saharan Africa<sup>[9]</sup>, especially on the left side of the heart<sup>[11]</sup>. While the vena-caval extension of WT into the right atrium is a well-documented phenomenon, left atrial involvement is extremely rare<sup>[11]</sup>. The frequency of such cases is presumably higher in this region where patients typically present with advanced disease, as seen in the case illustrated, hence the need for screening.

Screening for intravascular and intra-cardiac tumour extension can be done via CT scan, MRI, doppler ultrasound, or echocardiography<sup>[19]</sup>. Patients found to have intra-cardiac metastasis of WT should be referred to centers where appropriate interventions are feasible, as soon as the diagnosis is made.

Management of advanced WT is multi-disciplinary and multi-modal<sup>[10, 17]</sup>. Neoadjuvant chemotherapy has been proven to reduce the size of the atrioventricular tumour, with complete regression of the intra-cardiac mass in some reports<sup>[9, 10, 17-19]</sup>. Surgical management of intra-cardiac tumours is formidable and technically challenging, especially in LMICs<sup>[8-10, 18]</sup>. In drug-susceptible cases, pre-operative chemotherapy facilitates easy resection, thus minimizing surgical complications<sup>[7, 8, 18]</sup>. Urgent life-saving surgical intervention such as cardiopulmonary bypass is indicated in cases of imminent pulmonary embolism and affection of intrinsic cardiac function<sup>[5, 10]</sup>. The outcome of intravascular/intra-cardiac extension of Wilms tumour is favourable if detected early, and appropriate management promptly instituted<sup>[10, 19]</sup>.

### Conclusion and Recommendations

Left atrial involvement in WT is a rare but documented complication of the disease, and could be an incidental finding as it occurred in the index case. All patients with

WT should have echocardiography done as part of their routine evaluation to provide baseline information about cardiac function, before the possible commencement of potentially cardiotoxic medications and also to screen for intra-cardiac extension of the disease.

### References

1. Cunningham ME, Klug TD, Nuchtern JG, Chintagumpala MM, Venkatramani R, Lubega J, *et al.* Global disparities in Wilms tumor. *J. Surg. Res.* 2020; 247:34-51.
2. Hassan MA. Giant Wilms tumor in a child: Safe radical nephrectomy. *J. Pediatr. Surg. Case Reports.* 2020; 62:1-4.
3. Carter NH, Avery AH, Libes J, Lovvorn III HN, Hansen EN. Pediatric solid tumors in resource-constrained settings: A review of available evidence on management, outcomes, and barriers to care. *Children.* 2018; 5:1-9.
4. Mabadeje-Isowo AB, Sowunmi AC, Oyesegun AR, Adenipekun A, Ketiku KK, Okoro US. Management of Wilms tumor at the Lagos University Teaching Hospital (a 10-year retrospective study). *J. Clin. Sci.* 2019; 16:68-73.
5. Kajal K, Munirathinam GK, Mandal B, Gandhi K, Singh H, Kanojia R. Wilms tumor with intra-cardiac extension causing dynamic tricuspid valve obstruction: An anesthetic challenge. *Saudi J. Anaesth.* 2018; 12:321-323.
6. Apple A, Lovvorn III HN. Wilms Tumor in Sub-Saharan Africa: Molecular and social determinants of a global pediatric health disparity. *Front. Oncol.* 2020; 10:1-7.
7. Davidoff AM. Wilms' tumor. *Curr. Opin. Pediatr.* 2009; 21:357-364.
8. Altwaeel H, Kabbani MS, Al Shammari A, Al-Namshan M, Alghamdi AA. Perioperative management of Wilms tumor with intracardiac extension: Report of two cases with review of literature. *J. Saudi Heart. Assoc.* 2020; 32:106-109.
9. Mapelli M, Zagni P, Ferrara R, Calbi V, Mattavelli I, Muratori M, *et al.* Unexpected huge prevalence of intracardiac extension of Wilms tumor—a single center experience from a Ugandan hospital. *Children.* 2022; 9:743.
10. Qureshi SS, Bhagat M, Smriti V, Murli D, Baheti A, Yadav S, *et al.* Intravascular extension of Wilms tumor: Characteristics of tumor thrombus and their impact on outcomes. *J. Pediatr. Urol.* 2021; 17:69.e1-69.e1.
11. Sahu I, Ruhela M, Ola RK. Left atrial metastasis of a Wilms' tumor: A rare occurrence. *J Fam Med Prim Care.* 2020; 9:44604462.
12. Kuerten BG, Brotkin S, Bonner MJ, *et al.* Psychosocial burden of childhood sickle cell disease on caregivers in Kenya. *J Pediatr Psychol.* E-pub ahead of print, 2020. Doi: 10.1093/jpepsy/jsaa021.
13. Prasad M, Vora T, Agarwala S, *et al.* Management of Wilms tumor: ICMR consensus document. *Indian J Pediatr.* 2017; 84:437-445.
14. Wilde JCH, Laméris W, Van Hasselt EH, *et al.* Challenges and outcome of Wilms tumour management in a resource-constrained setting. *African J Paediatr Surg.* 2010; 7:159-162.
15. Elayadi M, Magdy S, Khalil E, *et al.* Management and

- outcome of pediatric metastatic Wilms' tumor at the National Cancer Institute, Egypt. *J Egypt Natl Canc Inst*; 32. E-pub ahead of print, 2020. Doi: 10.1186/s43046-020-00031-7
16. Israels T, Moreira C, Scanlan T, Molyneux L, Kampondeni S, Hesselting P, *et al.* SIOP PODC: Clinical guidelines for the management of children with Wilms tumour in a low-income setting. *Pediatr blood & cancer*. 2013; 60:5-11.
  17. Shamberger RC, Ritchey ML, Haase GM, *et al.* Intravascular extension of Wilms tumor. *Ann Surg* 2001; 234:116-21.
  18. Abdullah Y, Karpelowsky J, Davidson A, *et al.* Management of nine cases of Wilms' tumour with intracardiac extension - A single centre experience. *J Pediatr Surg*. 2013; 48:394-399.
  19. Schettini ST, Da Fonesca JHAP, Abib SCV, *et al.* Management of Wilms tumor with intracardiac extension. *Pediatr Surg Int*. 2000; 16:529-532.