

## CASE REPORT



# High-Risk Adult Wilms' Tumour in Pregnancy: A Case Report

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## Abstract

Wilms' tumour is the most common renal neoplasm in children with an incidence of 10 cases per 1 million children and a median age at diagnosis of 3.5 years. In Western countries its occurrence in adults is 0.2 cases per million people in western countries and carries a poorer prognosis. The co-existence of Adult Wilms' tumour and pregnancy is extremely rare with less than 20 cases published in the English literature. We present a case of a Malawian woman who had progressive high-risk metastatic Adult Wilms' tumour in pregnancy after nephrectomy, radiotherapy and two lines of chemotherapy.

**Keywords:** Africa, Antineoplastic Combined Chemotherapy Protocols, incidence, kidney, Kidney neoplasms, Malawi, pregnancy, Wilms' tumour

## Introduction

The coexistence of pregnancy and malignancy is a rare phenomenon with an incidence rate of 1 in 1000 pregnancies in the developed world<sup>1-4</sup>. The incidence in Africa is thought to be lower than that of developed countries because women in developing countries get pregnant at younger ages<sup>5</sup>. Among the malignancies that have been recorded to co-exist with pregnancy, 70-80% of the cases were the following cancers: breast, cervical, lymphoma, ovarian, and melanoma<sup>2,6</sup>.

Queen Elizabeth Central Hospital (QECH) records indicate that 11 (1.05%) of female cancer patients who presented between 1 January 2015 and 31 December 2017 were pregnant. Among these patients, haematological malignancies were the most common (n=6) followed by breast carcinoma (2), Kaposi's sarcoma (1), Osteosarcoma (1) and Wilms' tumour (1).

Wilms' tumour has an incidence of 10 cases/million children with a median age of 3.5 years at the time of diagnosis<sup>7,8</sup>. It accounts for 6% of paediatric malignancies and is associated with a 42% five-year event free survival at QECH<sup>9,10</sup>. Adult Wilms' Tumour (AWT) has an estimated incidence of <0.2 cases/million individuals in western countries (0.5% of all adult renal neoplasms)<sup>11-13</sup>. Its incidence is associated with mutations in WT1, CTNBN1, IGF2 and WTX genes<sup>14</sup>. Adult Wilm's tumour has a worse prognosis due to lack of established treatment protocols, its highly aggressive course and metastatic potential<sup>7,13</sup>. Adults also tend to have a lower drug toxicity threshold and advanced disease at the time of diagnosis<sup>7,13,15</sup>.

Adult Wilm's tumour diagnosis is based on the criteria developed by Kilton et. al which are: a primary renal neoplasm in the age group of >15 years with histological features of embryonic glomerulo-tubular structure with

immature spindle or round cell stroma and no areas of tumour diagnostic of renal cell carcinoma<sup>11,13</sup>.

## Case Presentation

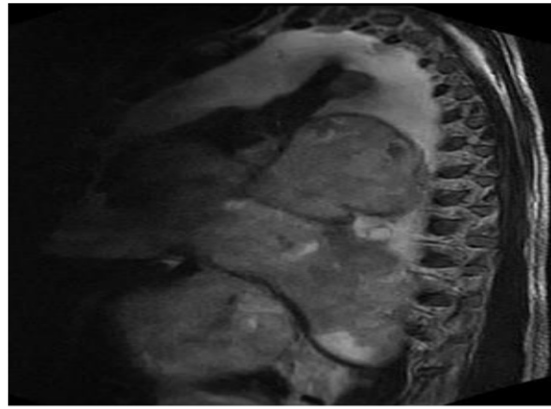
In January 2016, a 23 year old HIV negative woman presented at the QECH oncology clinic with a diagnosis of International Society of Paediatric Oncology stage IV High risk AWT made at Nottingham University Hospital (August 2014). At the time of diagnosis, she had a primary tumour in the left kidney, a solitary liver metastasis and multiple right lung metastases. During her treatment course in Nottingham, she had actinomycin, vincristine and doxorubicin induction chemotherapy (August-October 2014) with partial response. This was changed to high-risk protocol: cyclophosphamide/doxorubicin alternating with etoposide/carboplatin (November 2014).

In January 2015, she had a resection of the primary tumour and liver metastasis. Radiotherapy to the left flank to a dose of 24 Grays and the whole right lung to a dose of 18 Grays followed (February 2015) and Doxorubicin was omitted during her radiotherapy. In September 2015 upon completion of chemotherapy, CT scan confirmed partial response of her lung metastases and no evidence of disease below the diaphragm. Resection of the lung lesions was not feasible due to their multiplicity. Therefore close monitoring was opted for. No other significant medical history was available.

At the time of presentation at QECH, socio-economic circumstances had limited her oncology care options to QECH. She also had no radiological evidence of disease progression, an Eastern Cooperative Oncology Group performance status (ECOG) score of 1 (ambulatory but incapable of strenuous work) and normal laboratory results. After 15 months on follow-up, she developed intermittent dyspnoea and right-sided chest pain. Chest radiograph

revealed right lung fibrotic changes in the upper lobe with collapse of the middle and lower lung zone (Figure 2). A coincidental discovery of pregnancy (G2P1) with an intrauterine singleton pregnancy of 15 weeks gestation age by ultrasound scan was also made. After non-directive counselling she opted for close follow-up.

A foetal anomaly scan at 22 weeks gestation revealed no anomalies and subsequent growth scans were normal. She developed lower back pain and had elevated serum calcium (3.2mmol/L, normal range: 2.0-2.5 mmol/L), Alkaline phosphatase (128.2U/L; 0-126 U/L) and lactate dehydrogenase (754.3 U/L; 0-248 U/L). MRI showed: a large left pleural effusion with multiple heterogeneous masses within the left lung, large mediastinal lymph nodes, and a large heterogeneous right kidney mass (Figures 3 & 4). She was started on Zoledronate 4mg IV and had a miscarriage two days later at 25 weeks gestation. She presented again 10 days later and was started on chemotherapy while having an ECOG score of 3 (bedbound for >50% of waking hours), cachexia and pitting oedema. Throughout the follow-up period she maintained normal renal function and had no thromboembolic events. She died of lung failure two days after starting ifosfamide, carboplatin and etoposide.



**Figure 4: Sagittal T2 weighted MRI sequence**

## Discussion

We accessed 10 reports on the coexistence of AWT with pregnancy in published English literature<sup>16-26</sup>. It is our postulation that progressive tumours precipitated the miscarriage and this could have potentially been mitigated by early onset chemotherapy. Previous exposure to heavy treatment of her metastatic disease and declining performance status however portended a poor prognosis<sup>27</sup>.

Our patient also experienced the co-existence of several factors associated with the development of psychological distress namely: (I) young age, (II) advanced and recurrent disease, (III) declining functional status, (IV) previous experience with cancer therapy, (V) presence of socio-economic challenges and (VI) the maternal-foetal beneficence conflict resulting from cancer in pregnancy<sup>28-34</sup>. The presence of psychological distress has been associated with disease progression or relapse, reduced quality-of-life, impaired social relationships, increased risk of suicide, longer rehabilitation time, poor adherence to treatment and shorter survival<sup>30,35-37</sup>. It is for this reason that distress and psychological support have been designated as a sixth vital sign and human right in cancer patients respectively<sup>38,39</sup>.

Management of pregnancy-associated cancer is difficult due to lack of robust evidence, paucity of established treatment guidelines and complexity of considerations including beneficence obligations of clinicians to the mother and foetus<sup>28,40-43</sup>. Care for these patients in resource limited settings is also challenged due to availability of: (I) access to care, (II) diagnostic services, (III) treatment and (IV) personnel for interdisciplinary care<sup>10,39,44-47</sup>. Thus patients are likely to be diagnosed with advanced disease and experience delays in starting treatment; with consequential poor survival.

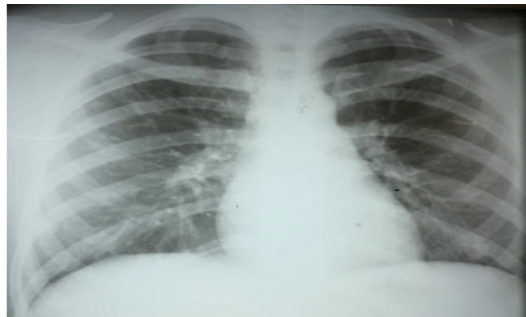
These challenges reflect resource distribution challenges which require long term investment in health care. The maternal-foetal ethical conflict highlights the importance of patient education on fertility issues and provision of contraception.

## Consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

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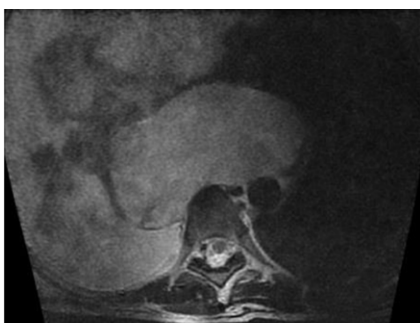
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**Figure 1: X-ray image from January 2016**



**Figure 2: X-ray image from March 2017**



**Figure 3: Axial T2 weighted MRI sequence**

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