

# Nephroblastoma with Spinal metastasis in a 12 year old Nigerian female: A case report

## Abstract

**Background:** Nephroblastoma, also known as Wilms' tumor, is the most common malignant renal tumor in childhood. Nephroblastoma is an embryonal tumor consisting of blastemal, stromal and epithelial elements. The median age for the diagnosis of nephroblastoma is 3.5 years. Occurrence beyond 5 years of age is rare. Treatment of nephroblastoma is multimodal.

**Case report:** We report an unusual case of Nephroblastoma with spinal metastasis in a 12 year old Nigerian female. The patient presented to us with 8 week history of left sided abdominal mass, 7 week history of abdominal pain and 6 day history of inability to walk, all prior to presentation. She had nephroureterectomy and chemotherapy. Histology of the specimen confirmed nephroblastoma and spinal magnetic resonance imaging showed spinal metastasis.

**Conclusion:** Nephroblastoma can occur in an adolescent with an uncommon site of metastasis to the spine.

**Keywords:** nephroblastoma, wilms, adolescent, metastasis, spine

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## Introduction

Nephroblastoma, also known as Wilms' tumour, is the most common malignant renal tumor in children accounting for about 95% of all pediatric tumors of the kidney. Overall, nephroblastoma is the fifth most common pediatric malignancy.<sup>1</sup> It is an embryonal tumor that develops from the remnant of persistent metanephric tissue and is made up of histologic elements namely blastemal, stromal and epithelial.<sup>1</sup> Nephroblastoma is relatively more common in blacks than in whites and is associated with a number of syndromes such as WAGR, Beckwith Wiedemann, and Denys-Drash syndromes.<sup>2</sup> Nephroblastoma usually present as a painless abdominal mass which may be associated with haematuria, hypertension, and fever. The peak age of incidence of nephroblastoma is 3.5 years. Incidence beyond 5 years of age is rare.<sup>3</sup> Prognosis of nephroblastoma depends on the tumor stage, biological factors and histological subtype. Treatment of nephroblastoma is multimodal which includes surgery, chemotherapy and/or radiotherapy.<sup>3</sup> We report an unusual case of nephroblastoma with spinal metastasis in a 12 year old Nigerian female. The focus of this article is to draw attention to this unusual case of nephroblastoma, which was painful, in a 12 year old and for clinicians to consider the diagnosis of nephroblastoma in older children that have renal masses.

## Case Report

Our patient is a 12 year old female who presented on account of 8 week history of left sided abdominal mass, 7 week history of abdominal pain and 6 day history of inability to walk, all prior to presentation. The abdominal mass was insidious in onset and progressively increased in size over time. One week after the onset of the abdominal mass, the patient developed abdominal pain which was located over the mass, dull in nature, non-radiating, aggravated

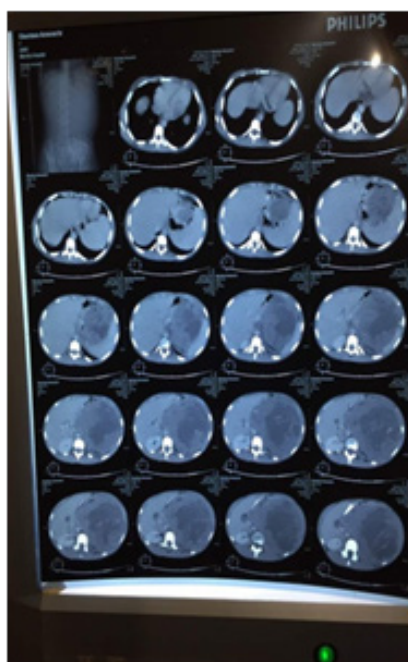
by activity and application of pressure over the mass. There was no associated vomiting or constipation. The patient subsequently developed inability to stand and could not walk. There was no history of preceding trauma.

On clinical evaluation, she is an adolescent female in painful distress, conscious, afebrile, mildly pale. Temperature was 36.7°C, respiratory rate was 34 cycles per minute, pulse rate was 90beats per minute, blood pressure 110/90mmHg. Abdominal examination revealed a mass located in the left lumbar region extending to the left iliac fossa, measuring 10cm by 12cm in its wildest diameter. This abdominal mass was firm, tender and did not cross the midline. There was no ascites (Figure 1). Neurological assessment revealed hypotonia on the lower limbs (normotonia on the upper limbs), grade 2 power on both lower limbs (grade 5 on both upper limbs) and hyporeflexia on both lower limbs (normoreflexia on the upper limbs).

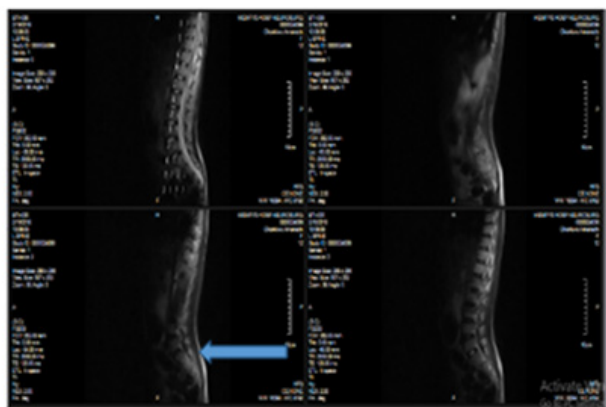


Figure 1 Clinical photograph of the index patient.

On investigation, urinalysis, complete blood count, serum electrolytes, urea and creatinine were normal. Abdominal ultrasound showed a left renal mass that was totally replaced by a complex partly cystic and partly solid mass, 20cm by 17cm in size. Computed tomography (CT) scan of the abdomen and pelvis showed a left renal mass with more cystic than solid components extending from the left hemidiaphragm to the left iliac fossa. The right kidney appears normal (Figure 2). Magnetic resonance imaging (MRI) of the spine showed evidence of spinal metastasis (Figure 3).



**Figure 2** CT scan of the index patient showing the left Wilms' tumour.



**Figure 3** Spinal MRI, arrow showing the site of spinal metastasis.

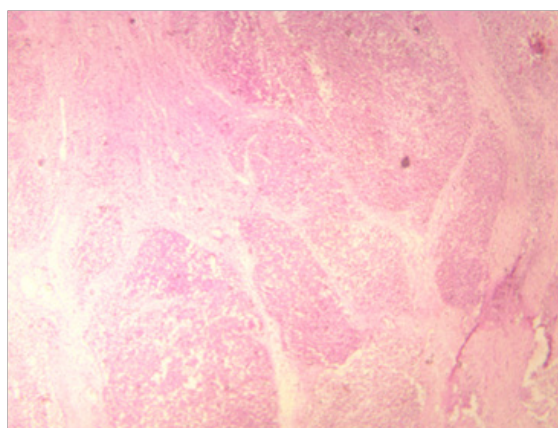
The parents were counseled and informed consent obtained. Patient was optimized and had left nephroureterectomy. The specimen (Figure 4) was sent for histology.

Histology result showed a triphasic tumour composed of sheets and nodules of undifferentiated blastemal cells, tubules and glomeruloid epithelial structure (Figure 5). The histological diagnosis was Wilms' tumour. She was placed on adjuvant chemotherapy which consisted of Vincristine, Actinomycin D, Adriamycin and Cyclophosphamide.

The patient could not get radiotherapy due to absence of the facility. Unfortunately, she was lost to follow up.



**Figure 4** Resected specimen in a kidney dish.



**Figure 5** Photomicrograph of a section of the specimen (H & E, X10 magnification).

## Discussion

Nephroblastoma (Wilms' tumour) was first described by Max Wilms in 1899. It is malignant renal tumours that account for 6% of all pediatric tumors and is associated with metastatic disease in 12% of children at diagnosis.<sup>4</sup> Nephroblastoma is usually diagnosed in children less than 5 years of age. Incidence of this tumor beyond 5 years of age is rare. Most reported cases of nephroblastoma are documented to have occurred between the ages of 3 and 4 years.<sup>2,5</sup> Nephroblastoma was diagnosed in the index patient at the age of 12 years.

Nephroblastoma is rare in adolescents and adults accounting for 0.5% of all renal tumors in adult and only 3% of Wilms' tumors are reported in adults.<sup>6</sup> The prognosis of nephroblastoma in adult is poor when compared with children. This is because adults present with advanced disease stage and chemotherapy have moderate effect.<sup>7</sup> The tumor stage and histopathology of the tumor are the most important prognostic factors.<sup>8</sup> Due to rarity of nephroblastoma in adolescents and adults, there is no established protocol for its treatment. Therefore,

adolescents with nephroblastoma are at a risk of either under treatment or incorrect treatment.<sup>9</sup> Currently, the staging and management protocol of both adult Wilms' tumor and paediatric Wilms' tumor are done in the same way, according to the National Wilms Tumor Study Group (NWTSG) and International Society of Paediatric Oncology (SIOP).<sup>7</sup> NWTGS advocates upfront nephrectomy to define the accurate stage of the tumor and the histology, on which further treatment stratification is decided. In contrast, the SIOP advocates the concept of pre-nephrectomy chemotherapy in all patients above 6 months of age to reduce the tumor size and prevent intraoperative tumor rupture causing spillage.<sup>10</sup> Nephroblastoma in adolescents and adults is associated with increased risk of recurrence.<sup>11</sup>

Nephroblastoma typically metastasizes to the lungs.<sup>1</sup> Metastasis to the central nervous system is possible but rare.<sup>7</sup> The index case has intraspinal metastasis. Ramdial et al gave an account of five Wilms' tumor adult patients that have paraplegia secondary to spinal metastasis. Four out of five patients died. One patient had neurological recovery following surgery though there was a relapse 4 months later.<sup>12</sup> In Wilms' tumor with spinal metastasis, the tumor may respond transiently to chemotherapy initially but there is always a re-growth of the tumor.<sup>13</sup> Spinal metastasis may be noticed few months after resection of Wilms' tumor.<sup>14</sup>

A literature search showed that there is no published report of Wilms' tumor with spinal metastasis in an adolescent in the West African sub region.

## Conclusion

Nephroblastoma that was hitherto known to occur in children less than 5 years of age, can occur in an adolescent in our environment. Metastasis from nephroblastoma can occur in an unusual site, the spine.

## Competing interest

The authors declare that they have no competing interests.

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None.

## Authors' contribution

All the Authors met the ICMJE criteria for authorship. All the authors took part in the concept, design, drafting of the manuscript and final approval.

## Informed consent

Consent was obtained from the patient's parents.

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