East African Medical Journal Vol. 96 No. 1 January 2019
ADULT WILMS' TUMOUR IN PREGNANCY
Dr. Owen Mang'usu, MBChB, Resident, Department of Surgery, Jaramogi Oginga Odinga Teaching and Referral Hospital, P. O. Box 3742-40100 Kisumu, Dr. Bryan Amata, MBChB, Medical Officer, Department of Surgery, Jaramogi Oginga Odinga Teaching and Referral Hospital, Kisumu, P. O. Box 12427-00100 Nairobi, Dr. Cindy Oyaya, MBChB, Medical Officer, Department of Surgery, Jaramogi Oginga Odinga Teaching and Referral Hospital, Kisumu P. O. Box 4077-00506 Nairobi, Dr. Caesar Bitta, MD, MMed (General Surgery), Lecturer, Department of Surgery Maseno University, Consultant Surgeon, Jaramogi Oginga Odinga Teaching and Referral Hospital, Kisumu Private Bag, Maseno, Dr. Francis Odira, MBChB, MMed (General Surgery), Lecturer, Department of Surgery, Uzima University College, Consultant Surgeon, Jaramogi Oginga Odinga Teaching and Referral Hospital, P.O. Box 19072-40100 Kisumu, Dr. Mbayah Etabale, MBChB, MMed (Pathology) Lecturer, Department of Pathology, Maseno University, Private Bag Maseno.

Corresponding author: Dr. Caesar Bitta, MD, MMed (General Surgery), Lecturer, Department of Surgery Maseno University, Consultant Surgeon, Jaramogi Oginga Odinga Teaching and Referral Hospital, Kisumu. Email; cbittas@maseno.ac.ke

ADULT WILMS' TUMOUR IN PREGNANCY

O. Mang'usu, B. Amata, C. Oyaya, C. Bitta, F. Odira and M. Etabale

ABSTRACT

Wilms' tumor (nephroblastoma) is the most common primary renal malignancy in children, with a peak presentation in the ages 3-4 years. Wilms' tumor is extremely rare in adults with around 300 cases described in literature. This presents both a diagnostic and treatment challenge in adults due to lack of standard protocols. Herein, we report a case of a 28-year-old primigravida who underwent nephrectomy postpartum for presumed renal cell carcinoma that was diagnosed as Wilms' tumor at histology. This case is being reported on account of its peculiar presentation, and its presentation in pregnancy, to provoke further research and thus improve management in the adult patient.

INTRODUCTION

Wilms' tumor (nephroblastoma) is the most common primary renal malignancy in childhood, mostly presenting by age 5 (2,3). Wilms' tumor is extremely rare in adults (4,5) and is even more uncommon in pregnancy (5,6). Due to lack of characteristic clinical and/or radiological presentation (5,7), it is diagnosed at histology following nephrectomy for presumed renal cell carcinoma which is much more common in adulthood (8,9). It thus presents as both a diagnostic and treatment challenge in adults, with management relying on existing protocols for children (1,10). It is for this reason that we report a case of a 28-year-old primigravida who underwent nephrectomy in the immediate postpartum period for presumed renal cell carcinoma, that at histology was confirmed to be Wilms' tumor. On account of its peculiar presentation, and its presentation in pregnancy, we present this case to provoke further research and thus improve management in the adult patient.

CASE PRESENTATION

A 28-year-old primigravida at 33 weeks gestation presented to her antenatal clinic with a 2-day history of bilateral lower limb swelling, facial puffiness and epigastric pain. Her past medical, and family and social history were unremarkable. On examination she was noted to have facial puffiness, bipedal edema and an elevated blood pressure of 163/113 mmHg. Systemic examination was unremarkable except for a gravid abdomen with a regular fetal heart rate. Urinalysis revealed red cells (4+) with proteinuria (3+) and her renal function tests were remarkable for mild elevation in creatinine (96 μmol/L against a reference of 39-91 μmol/L). She was subsequently referred for admission with a working diagnosis of severe pre–eclampsia and started on Methyldopa and Nifedipine.

Induction of labor was attempted unsuccessfully necessitating caesarean delivery 2 days after admission. Intra-operatively, following successful delivery of the fetus, the patient was noted to have a left upper quadrant retroperitoneal mass that was not explored further. Post-operatively, she had better control of her blood pressure on the antihypertensives. However, she was noted to have persistent left flank pain with minimal relief on analgesia. On examination she had a bimanually palpable, firm and tender left upper quadrant mass that on ultrasound was identified as a homogenous mass lesion on the left kidney. A CT scan of the abdomen revealed a large well-defined soft tissue mass arising from the left kidney that measured 21.8x20.6x14.7cm and showed poor heterogenous contrast enhancement. There were no radiological signs of metastasis. A hemogram done noted normocytic normochromic anemia with a hemoglobin of 6.5g/dl.

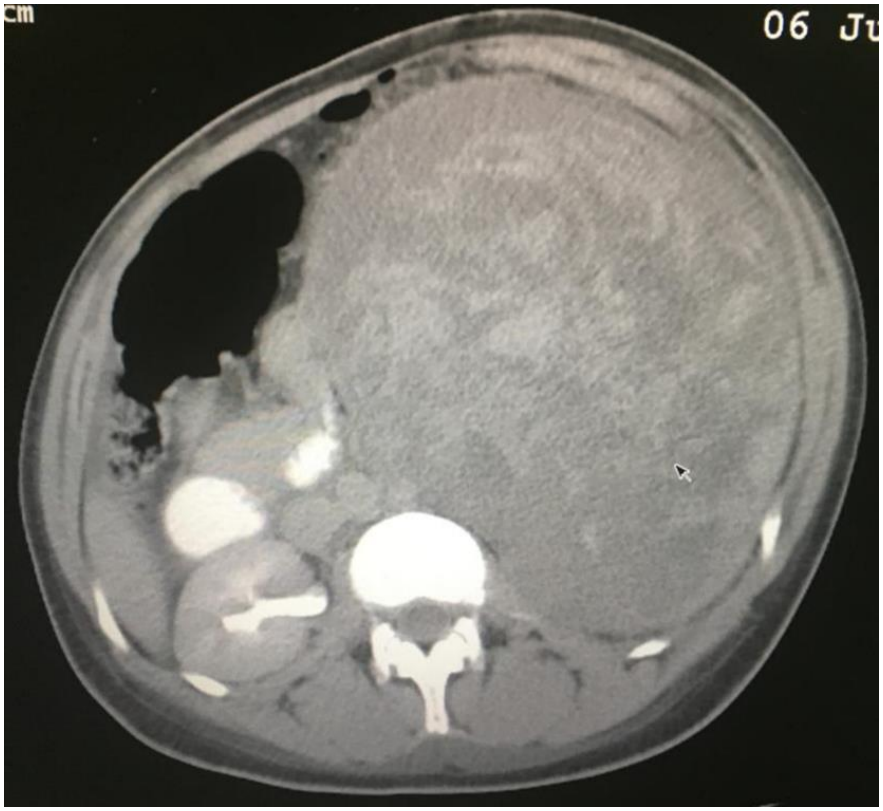

Figure 1- Axial CT scan of the abdomen showing a soft tissue mass arising from the left kidney

A diagnosis of probable renal cell carcinoma was entertained, and the patient was optimized for surgery. Six (6) days post caesarean delivery she underwent left radical nephrectomy via trans-peritoneal approach; no overt signs of metastasis were noted. She was discharged with her newborn ten (10) days later after an uneventful post-operative period.

At histology, the nephrectomy specimen measured 210 x 190 x 120mm and weighed 2345g. It had prominent vascularity on the external surface and cut sections showed fleshy encephaloid lobulated architecture with marked haemorrhoids and necrosis. There was an eccentrically located residual kidney 70 x 40mm with a clearly defined capsule, cortex and medulla. No intramural thrombi were noted within the hilar blood vessels and the ureter was unremarkable. Microscopy revealed a well circumscribed but unencapsulated tumor comprising of abundant undifferentiated blastema, diffuse pattern. Tumor cells were densely packed primitive small blue cells with scanty cytoplasm and overlapping nuclei with finely dispersed chromatin, scanty fibroblast-like stroma and epithelial elements that comprise of abortive tubules. There was no heterologous epithelial and stromal elements, no anaplasia, and no tumour was seen in the capsule or perirenal fat. Morphologic features were thus noted to be consistent with Wilms' tumour (stage pT2).

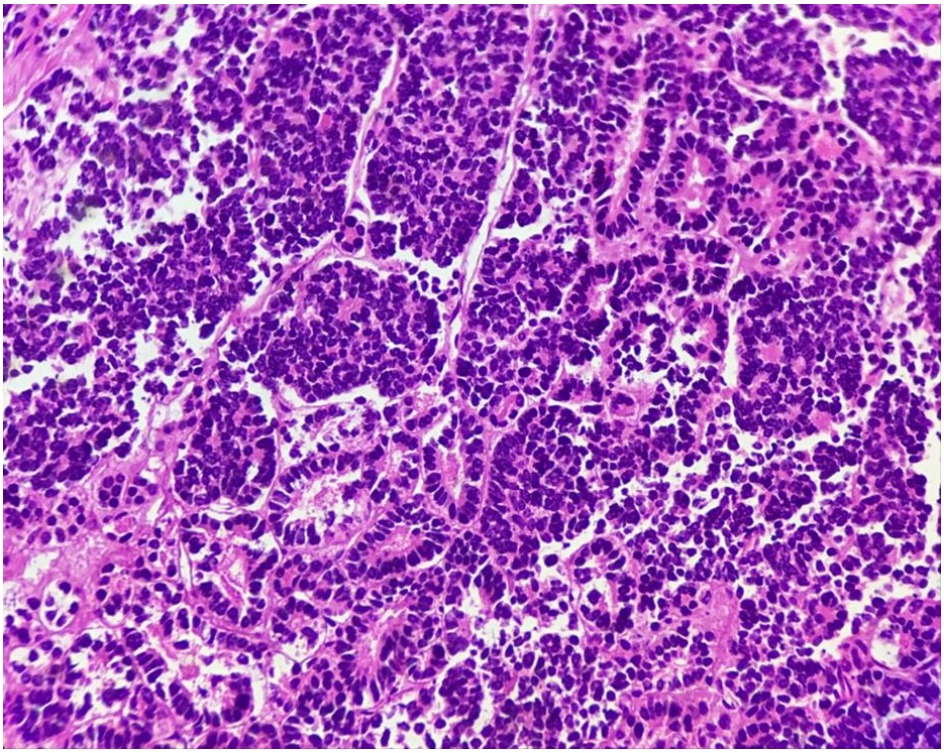

Figure 2 - The neoplasm is composed of small, round-to-oval cells with hyperchromatic nuclei and scanty cytoplasm. The blastema component is composed of cells forming solid clusters whereas the primitive epithelial component is composed of cells forming tubular structures

The patient was subsequently referred for oncological review and further management but opted not to have chemotherapy. She remains on surgical follow-up.

DISCUSSION

Wilms' tumour is the most common primary malignant renal tumour in children, and one of the commonest childhood solid organ tumours[1,2,3]. It accounts for about 6% of paediatric malignancies with a median age at presentation 3 to 4 years[3]; 90% of children are diagnosed before the age of 7 years[4]. A population-based registry study on childhood cancers (up to 19 years of age) across different countries and economies noted that renal tumours are more common in children younger than 5 years with decreasing relative frequency in the older age groups. Including other embryonal tumours, Wilms' tumour occurs primarily in children[3]. It is extremely rare in adolescents and adults with up to 300 cases reported in literature as of 2004[4]. Furthermore, there are very limited cases of Wilms' tumor in pregnancy with only 9 cases reported to date[5,6].

Wilms' tumor arises from abnormal proliferation of metanephric blastema without differentiation into glomeruli and tubules[8]. At histology there is no difference between adult and childhood Wilms' tumor; both characteristically have triphasic histology with variable components of blastemal, epithelial, and stromal structures[2]. As was in our case, blastemal component is usually predominant in adult Wilms' tumor, and is associated with more aggressive disease[9].

As in our case, the diagnosis of Wilms' tumor in adults is usually unexpected being confirmed at histology following nephrectomy for suspected renal cell carcinoma which is much more common in adults[8,9]. Preoperatively, diagnosis is difficult as there is no typical clinical presentation or radiographic signs that distinguish it from the more common renal cell carcinoma[5]. Core needle biopsy has been discouraged in some literature as wrong diagnosis is likely due to patchy distribution of the components[7].

Criteria for diagnosis of adult Wilms' tumour was defined by Kilton et al:

a) Primary renal neoplasm
b) Primitive blastematous spindle or round cell component
c) Formation of abortive or embryonal tubular or glomeruloid structures
d) No area of tumour diagnostic of hypernephroma (renal cell carcinoma)
e) Pictorial confirmation of histology
f) Age > 15 years

As in our case, adult patients tend to present with flank pain and hematuria as opposed to palpable boggy abdominal mass in children[7,9]. Pain has been described as the most common presentation in pregnancy[6]. Adult patients also tend to present with more advanced disease with worse prognosis[9].

As adult Wilms' tumour is rare, randomised trials cannot be conducted. Consequently, results of randomised trials with childhood Wilms' tumour have been extrapolated, with paediatric treatment guidelines being used in adult patients[1,8]. These are the National Wilms' Tumour Study Group (NWTSG), and the International Society of Pediatric Oncology (SIOP) protocols. NWTSG recommends primary surgery before administration of chemotherapy; SIOP advocates administration of four weeks of chemotherapy prior to surgery[2]. Wilms' tumour is both chemo- and radiosensitive and treatment is invariably multimodal[1,12]. These multidisciplinary management models have resulted in improved survival in children from less than 30% to more than 90%[2]. This has been proposed to show promise when applied to adult patients[8,10].

Radical nephrectomy is the standard of care for resectable disease, with the transperitoneal route preferred to provide adequate exposure for complete staging[2]. This is combined with chemotherapy with or without radiotherapy depending on stage of the disease.

Long-term follow-up is crucial to monitor for recurrence and drug toxicity[12]. Despite opting not to have further treatment, our patient remains on surgical follow-up.

CONCLUSION

Wilms' tumor is both a diagnostic and treatment challenge in adults, with management relying on existing protocols for children. On account of its peculiar presentation, and its presentation in pregnancy, we present this case to provoke further research and thus improve management in the adult patient.

REFERENCES

1. Geethamani V., Kusuma V., Gowda K.M.S., Saini M.L. Adult Wilms' tumour: a case report with review of literature. *Diagnostic Pathology* 2006, 1:46 https://doi.org/10.1186/1746-1596-1-46
2. Tongaonkar H.B., Qureshi S.S., Kurkure P.A., et al. Wilms' tumour: An update. *Indian J Urol.* 2007 Oct-Dec; 23(4):458-466. https://doi.org/10.4103/0970-1591.36722
3. Faranoush M., Bahoush G.R., Mehrvar A., et al. Wilm's Tumour: Epidemiology and Survival. *Research Journal of Biological Sciences* 4 (1): 86-89, 2009.
4. Madan A., Sonpavde G. (2016) 'Adult Wilms' Tumour', in Pagliaro L. (ed.). *Rare Genitourinary Tumours*. Switzerland: Springer International Publishing, pp. 79-93 https://doi.org/10.1007/978-3-319-30046-7_5
5. Khan I. A., Basu S., Khan D., et al. Wilms' Tumour in an Adult Female Complicating Pregnancy: A Case Report. *Journal of Clinical and Diagnostic Research*. 2018 June, Vol-12(6): PD05-PD06 https://doi.org/10.7860/JCDR/2018/32860.11658
6. Rehman J., Chughtai B., Guru K., Khan S.A., Adler H.L., Miller F. Wilm's tumor during pregnancy: report of laparoscopic removal and review of literature. *The Canadian Journal of Urology*. 2008 Aug;15(4):4180-3.
7. Pidakala P.L., Chaitra B., Pav K., Tejeswini V. Adult Wilms Tumour Cases Masquerading as Renal Cell Carcinoma. *National Journal of Laboratory Medicine.* 2018 Apr, Vol-7(2) PS01-PS04.
8. Szychot E., Apps J., Pritchard-Jones K. Wilms' tumour: biology, diagnosis and treatment. *TranslPediatr* 2014;3(1):12-24 https://doi.org/10.3978/j.issn.2224-4336.2014.01.09
9. Varma A.V., Malukani K., Rihal P., Nandedkar S.S. Adult Wilms' tumour: A case report with review of literature. *JCan Res Ther*. 2015;11:934-6.
10. Lenon M.S.L., Lopez R.A., Lusaya D.G. Adult Nephroblastoma (Wilms Tumour): A Case Report. *Med Surg Urol*.4: 150. https://doi.org/10.4172/2168-9857.1000150
11. Steliarova-Foucher E., Colombet M., Ries L.A.G., Moreno F., et al. International incidence of childhood cancer, 2001–10: a population-based registry study. *The Lancet Oncology*. Volume 18, Issue 6, p719-731 June 2017. https://doi.org/10.1016/S1470-2045(17)30186-9
12. Modi S., Tiang K.W., Inglis P., Collins S. Adult Wilms' Tumour: Case Report and Review of Literature. *J Kidney CancerVHL*. 2016;3(2):1-7. http://dx.doi.org/10.15586/jkcvhl.2016.52
13. Kilton L., Matthews M.J., Cohen M.H. Adult Wilms Tumour: A Report Of Prolonged Survival And Review Of Literature. *The Journal of Urology*. July 1980 Vol. 124:1–5. https://doi.org/10.1016/S0022-5347(17)55264-7