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Management of Wilms' Tumor in a Horseshoe Kidney at the Time of COVID-19 Pandemic: A Case Report

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Wilms' tumor is one of the most frequent tumors in childhood. The incidence of Wilms' tumor in a horseshoe kidney is estimated at 0.4 to 0.9%. It is highly treatable but a fast growing tumor. Management of cancer patients has become a dilemma for surgeons, due to the impact of the COVID-19 pandemic on the health care sector globally. Reported here is a rare case of a 2 year old boy, diagnosed with Wilms' tumor in a horseshoe kidney. This report aimed to highlight the multimodality treatment of Wilms' tumor and the modifications to cancer diagnostic and treatment protocols to adapt to the current health care crisis brought about by the pandemic.

Key words: COVID-19 pandemic, Wilms' tumor, horseshoe kidney

Wilms' tumor otherwise known as Nephroblastoma is the most common renal tumor in children.[1] Horseshoe kidney is the most common fusion anomaly of the kidneys. Its incidence varies from 1 in 400 to 1 in 1800 autopsies and is more common among males.[2] A report From the National Wilms' Tumor Study Group (NWTSG) done by Neville, et al. 2002, suggests that Wilms' tumor is 1.96 times more common in patients with horseshoe kidney than in the general population.[3]

Wilms' tumor is highly curable with survival rates as high as 90% as shown by the International Society of Pediatric Oncology-Renal Tumor Study Group (SIOP-RTSG) and the Children's Oncology Group (COG). However, Wilms' tumor is a fast-growing tumor with a doubling time between 2 – 3 weeks.[4] A comparison between UK and Germany showed that a delay in diagnosis between first symptoms and tumor diagnosis should be avoided.[5]

The COVID-19 pandemic had an unprecedented adverse impact on the spectrum of cancer care worldwide.[6] Majority of the health care workers and resources were allocated to address overwhelming number of COVID-19 patients. Locally, elective cases, including cancer surgeries, were put on hold as advised by the Philippine College of Surgeons. However, most childhood cancers behave aggressively and a delay in management may be detrimental. Modifications in cancer diagnostic and treatment protocols have been made to adapt to the current health care crisis brought about by the pandemic.

Presented here is a rare case of Wilms' tumor in a horseshoe kidney in a 2-year-old boy, managed successfully during the COVID-19 pandemic following a modified SIOP-RTSG 2016 protocol.

The Case

A 2-year-old Filipino male was admitted at the National Children's Hospital (NCH) for an asymptomatic kidney mass noted on abdominal CT scan. The imaging study was done from a referring institution to locate the undescended testes at 1 year of age, and showed bilateral undescended testes were located at the intraabdominal area, and an incidental finding of a renal mass in a horseshoe kidney. The patient was lost to follow-up from the previous referring institution during the start of COVID-19 pandemic.

Patient had been in excellent condition prior to admission. No known allergies, no history of hospitalizations nor previous surgery. No family history of malignancy.

The child was well-developed, well-nourished, ambulatory and not in distress. Pulse was 96 beats per minute, temperature 36.7ºC, and respirations 22 cycles per minute. Abnormal findings were limited to the inguinoscrotal area which revealed bilateral undescended testes. No abdominal masses were palpated.

The results of routine blood panel and urine analysis were within normal limits. The urinary excretion of vanillylmandelic acid, and catecholamines were also requested with normal results. An abdominal computed tomography (CT) scan confirmed a heterogeneously enhancing mass located in the inferior pole of left kidney measuring 3.0 cm x 2.8 cm x 2.8 cm, with an impression of horseshoe kidney with a mass in the left side (to consider Wilms' Tumor). No evidence of metastatic disease in the abdomen at this time. Chest CT scan with IV contrast showed no evidence of metastatic disease in the chest.

A multidisciplinary conference with Pediatric Oncology, Pathology, Pediatric Surgery, Radiology, and Pediatric Anesthesiology was conducted and the consensus was to follow the International Society of Pediatric Oncology (SIOP) guideline which recommends preoperative chemotherapy for all patients after diagnosis followed by nephrectomy.

The patient underwent 4 cycles of neoadjuvant chemotherapy (Vincristine + Dactinomycin). Repeat abdominal computed tomography (CT) done revealed stable left renal mass measuring 3.8 cm x 2.6 cm x 3.0 cm.

The patient, with his mother returned to the Out-patient Department and decided to proceed with the surgery. However, due to the COVID-19 pandemic, all elective surgeries were temporarily halted. Another multidisciplinary conference was conducted, and the decision was to continue neoadjuvant treatment, with a total of 16 cycles of chemotherapy (Vincristine + Dactinomycin).

The patient's condition was stable all throughout the neoadjuvant therapy. Repeat work-up done showed no evidence of metastases (Figure 1). The patient underwent surgery on October 28, 2020.

The surgery was performed by a team composed of a Pediatric Surgery consultant and Fellows in-training.

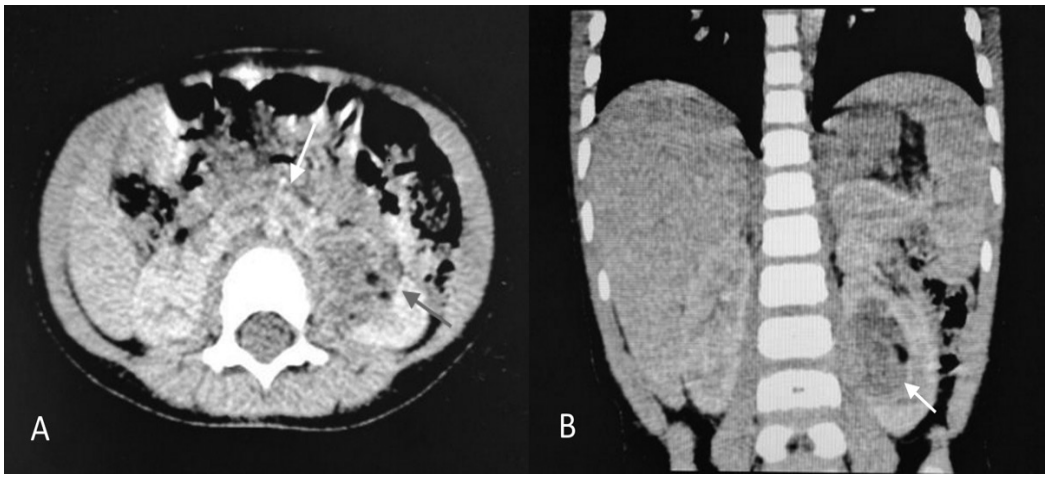

Figure 1. A. Axial CT scan of the abdomen at the level of L3 and L4, demonstrating the mass (gray arrow) and the inferior poles of both kidneys medially directed and connected by a homogenously enhancing parenchymal isthmus (white arrow) located anterior to the abdominal aorta and inferior vena cava. B. Coronal CT scan of the abdomen demonstrating a heterogeneously enhancing mass in the interpolar region and inferior pole of the left kidney, measuring approximately 3.1 cm x 2.9 cm x 2.9 cm. A good plane of demarcation (white arrow) is seen between the said mass and partially compressed middle and inferior infundibulum and pelvis of the left kidney.

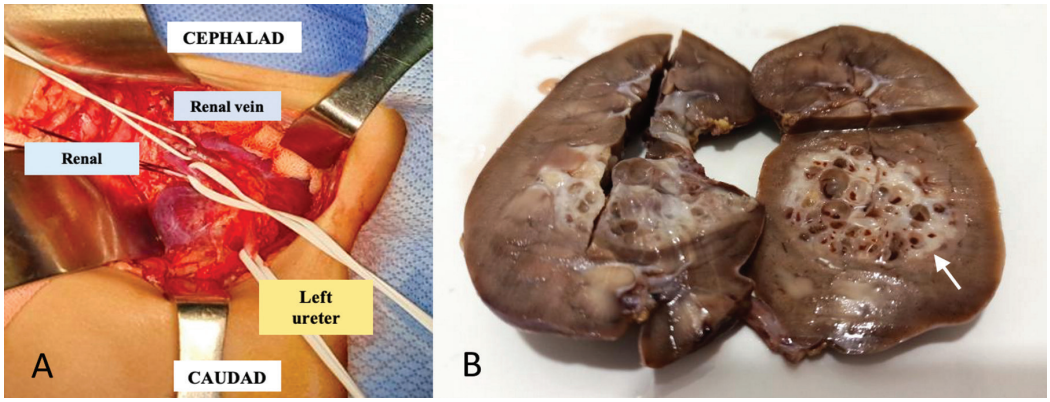

Figure 2. A. Tumor in situ. B. The left kidney was 64 g, measured 10 cm x 9 cm x 5.5 cm with an intact capsule and no attached perirenal fat. Cut section showed a 3.5 cm x 2 cm x 3 cm multicystic mass (white arrow) located at the middle to lower pole with no involvement of the renal pelvis. The locules measures from 0.2 cm to 0.6 cm in diameter and contained clear thin fluid.

The patient's abdomen was explored via left subcostal incision under general anesthesia. A solid, hard tumor with cystic component was found arising from the lower pole of the left segment of a horseshoe kidney with no infiltration of the surrounding structures (Figure 2). There was no note of metastatic spread on the paraaortic lymph nodes and the inferior vena cava. The rest of the intraabdominal structures were normal. The left renal artery was occluded using vessel loop and the demarcation of its supplied parenchyma was identified. The tumor was resected including the left renal artery, vein and ureter. The isthmus was divided with a harmonic scalpel and suture ligated using Vicryl 3-0 round. No other associated renal anomalies were noted. The left segment of the horseshoe kidney was supplied by a single renal artery and drain by a single renal vein. The left ureter was also identified. The total operating time was 205 minutes with estimated blood loss of 220 ml. The bilateral undescended testes were located high intrapelvic. Post-operative course was uneventful. The patient stayed at the surgery ward and was then discharged after 3 days.

Patient was initially seen thru telemedicine and advised face to face consult at the out-patient department 1 week after discharge and was doing well. The plan is to do a staged laparoscopic Fowler-Stephen procedure at 3 years of age for the bilateral undescended testes. Biannual imaging follow-up for the first 2 years was also advised, as most recurrences occur within that time period.

Microsections of the mass (Figure 3) revealed a residual neoplasm composed of predominantly cystic structures interspersed with neoplastic cells disposed in round tubules, nest and rosette like structures occupying less than 10% of the tumor bed. The findings were consistent with Nephroblastoma (Wilms' Tumor) of favorable histology, SIOP Histologic risk classification: Intermediate risk, Regressive type. Resection margins were free of tumor. The tumor was limited to the kidney and completely resected, SIOP stage 1. The patient was advised to have a surveillance CT scan after 6 months to monitor possible recurrence on the contralateral side.

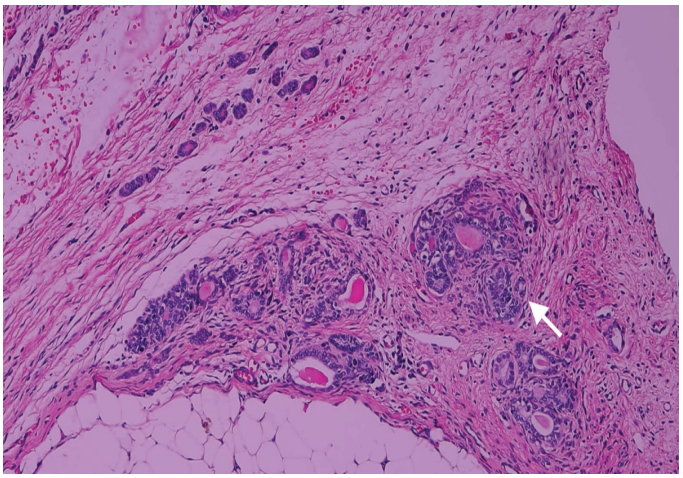

Figure 3. High power view (40x magnification) showing pseudo rosettes (arrow) on HE stain.

Discussion

Wilms' tumor otherwise known as Nephroblastoma is the most common renal tumor in children accounting for 87% of all renal mass and represents 7% of all malignant tumors in children.[1] It is known to occur in association with various syndromes, genetic and anatomical abnormalities especially those of the genitourinary tract. This includes genitourinary anomalies hypospadias, cryptorchidism and renal fusion anomalies. A report from the National Wilms' Tumor Study Group (NWTSG) done by Neville, et al. 2002, suggests that Wilms' tumor is 1.96 times more common in patients with horseshoe kidney than in the general population.[3]

Horseshoe kidney is the most common fusion anomaly of the kidneys. The incidence varies from 1 in 400 to 1 in 1800 autopsies and is more common among males.[2] Horseshoe kidneys most likely result from the union of the metanephros during the 4th and 6th week of gestation, before renal ascent.[8] The tumor usually involves the isthmus, but can be found in any region of the kidneys. In this case, the tumor involved the mid to lower pole of the left kidney. Although the etiology of Wilms' tumors in horseshoe kidneys has not yet been established, it has been proposed that the teratogenic factors that influence metanephric blastema to migrate and form the isthmus, may also be responsible for oncogenesis.[9]

Patients with Wilms' tumor in a horseshoe kidney can sometimes be very difficult to diagnose and must be approached in a meticulous manner. Most cases are asymptomatic in presentation and some are diagnosed incidentally as seen in this case. Accurate preoperative diagnosis is important for planning treatment modalities and may help decrease intraoperative complications. Abdominal CT scan is a reliable method for diagnosis. A study done by Jhobta, et al., suggests that a preoperative arteriography can be done to help identify any vascular anomalies associated with these types of cases although it is not routinely done.[10]

Multimodality treatment has been used to treat Wilms' tumor.[11] There are two main protocols in the current management of Wilms' tumor: The National Wilms' Tumor Study Group (NWTSG), which is being followed by North American Surgeons which is the upfront surgery principle, and SIOP Group, from Europe, which believes that upfront chemotherapy principle in all the stages of the disease is the optimal treatment for Wilms' tumor. Large numbers of patients have been studied in the clinical trials of both SIOP and NWTSG. Both approaches yield almost equivalent outcomes, 2 and 4 year overall survival rates stage for stage when their results were compared, though debate continues to discuss merits of each approach.[12-13]

The lockdown due to the COVID-19 pandemic was unprecedented. Adapting to the current health care situation to meet the needs of cancer patients was warranted. Surgical procedures were fraught with the risk of transmitting disease to health workers as well as patients most in danger of becoming severely-ill from the disease.

The management of the cancer patient has become a dilemma for surgeons. While most cancer surgeries are elective cases, a delay in diagnosis and treatment can have a deleterious effect on a patient's outcome. In order to overcome these challenges, the Philippine Society of Pediatric Surgeons proposed guidelines for handling patients waiting for cancer surgeries. The goal was to provide appropriate care to children with emergent and urgent surgical conditions while optimizing limited hospital resources and ensuring safety of all healthcare workers.[14] They recommended that urgent cases, which include most of cancer surgeries be given priority because any delay in these procedures could be detrimental.[15]

Sullivan, et al., had coordinated with leaders from SIOP- Europe, COG and SIOP-PODC (Pediatric Oncology in Developing Countries) to build consensus recommendations for the diagnosis and treatment of the six most curable cancers identified in the WHO Global Initiative for Childhood Cancer. This included Wilms' tumor. They provided practical advice for adapting diagnostic and treatment protocols for children with cancer during the pandemic, the measures taken to contain it (e.g. extreme social distancing) and how to prepare for the anticipated recovery period.[7]

Surgery and adjuvant therapy should be timed according to the protocol. Due to the delays caused by the pandemic, Sullivan, et al. recommended that if a patient had responded to chemotherapy, further courses

of preoperative chemotherapy should be continued until surgery could be performed. In case a tumor progressed despite preoperative chemotherapy, immediate surgery would be attempted despite the pandemic situation.[7]

In NCH, the authors follow the SIOP-RTSG 2016 protocol of pre-operative chemotherapy followed by surgery. Advocates of neoadjuvant therapy suggest that there is a greater chance of tumor resectability and less incidence of tumor spillage, mortality and morbidity.[16]

However, due to the indefinite suspension of elective surgery during the COVID-19 lockdown, modifications to the SIOP-RTSG 2016 protocol were made to adapt to pandemic health service restrictions. For this patient, a multidisciplinary conference was convened. The consensus was to proceed with the chemotherapy because no elective surgery was allowed at the time. The 16 cycles of chemotherapy (Vincristine + Dactinomycin) were completed over 6 months before elective surgery was performed.

A left subcostal incision can entirely provide access to a horseshoe kidney while providing adequate vascular control.[17] Horseshoe kidneys are usually positioned lower within the abdomen. The isthmus typically lies just in front of the aorta and IVC, at the level of the 3rd to 5th lumbar vertebral bodies. The location of the pelvicalyceal system is of utmost importance and should be identified during surgery. The most frequent complications from horseshoe kidney surgeries are urine leaks and ureteric injuries.[18]

Patients with a solitary tumor in a horseshoe kidney are managed with the same protocol used for a unilateral Wilms' tumor. Complete tumor resection with clear margins is the gold standard of treatment. It is imperative that the isthmus is resected. If the urine does not drain through the remaining kidney, a urinary fistula may result.[11]

Although the management of Wilms' tumor in a horseshoe kidney may prove to be very challenging, the overall survival of patients with Wilms' tumor in a horseshoe kidney appears to be about the same as for Wilms' tumor in normal kidneys.[19] The estimated survival 4 years after diagnosis was 86% in the NWTSG report.[3]

Conclusions

Wilms' tumor in a horseshoe kidney is a rare finding. Management can be difficult and even more so due to the challenges posed by the emergence of the COVID-19 pandemic. Wilms' tumor is a highly treatable but a fast growing tumor. Hence, cancer treatment should be given priority because any delay may affect the patients' chances of survival. Treatment planning should be made in the context of current COVID-19 pandemic. Modifications of existing protocols are necessary to adapt to the current health care situation to be able to provide oncologically appropriate treatment without compromising the safety of patients and the health providers.

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