Case Report

Wilms tumour in a two day old neonate: A rare and challenging case

Naveen Ravel[1], Teena Rajan[2], K Mohankumar[3], Gomathy Vadivelu[4], P N Sathiyamoorthy[5], Arun Ramanan[6], P Vijayaraghavan[7], Sam M Joel[7], Dheeraj Chodavarapu[2], Meena Konduri[2], Vijayalakshmi Pothala[2]

From [1]Professor and Head, [2]Senior Resident, [5]Associate Professor, [6]Assistant Professor, Department of Medical Oncology, [3]Professor and Head, [4]Associate Professor, [7]Assistant Professor, Department of Pediatric Surgery, Stanley Medical College, Chennai, Tamil Nadu, India

ABSTRACT

Wilms tumor or nephroblastoma is the second most common renal tumor occurring in the neonatal age group next to congenital mesoblastic nephroma. The most common age group of presentation of Wilms tumor is between 3 and 4 years of age. Here, we report a case of right-sided Wilms tumor in a 2-day-old neonate. A term-born female baby presented on day 2 of birth with abdominal distension and physical examination revealed a large palpable mass in the right lumbar region crossing the midline. Imaging features were suggestive of a retroperitoneal mass of probable right renal origin. The patient was taken up for right radical nephroureterectomy and Per-operatively, there was anticipated tumor spillage. Post-operative histopathological examination was suggestive of a triphasic Wilms tumor with no anaplasia and favorable histology. The patient was diagnosed with Wilms tumor of the right kidney, COG stage III, SIOP ("International Society of Pediatric Oncology") intermediate risk. Loss of heterozygosity testing for 1p and 16q was negative. She was started on chemotherapy with a DDA4 regimen. She completed whole abdominal radiotherapy and tolerated treatment well. The child is disease free and is under regular follow-up. This case of neonatal Wilms tumor was indeed a challenge to the pediatric surgeons and the oncologists but the final results were worth the challenge that was faced.

Key words: Chemotherapy, Neonatal tumors, Radiotherapy, Wilms tumor

Wilms tumor or nephroblastoma is the second most common renal tumor occurring in the neonatal age group next to congenital mesoblastic nephroma (CMN) [1]. The most common age of presentation of Wilms tumor is 3–4 years [2]. Treatment guidelines are well established for the management of Wilms tumor as per the National Wilms Tumor Study group (NWTS) and the International Society of Pediatric Oncology (SIOP) protocols. The survival rate is >90% with combined modality treatment [3].

Since only a very few handful of neonatal Wilms tumor cases have been reported [4], we report a case of right-sided Wilms tumor in a 2-day-old neonate.

CASE REPORT

A term female baby, born to a 21-year-old mother by a normal full-term vaginal delivery with an uneventful antenatal period, on day 2 of birth was referred to us with complaints of abdominal distension and mass per abdomen since birth.

Access this article online	
Received - 14 November 2022 Initial Review - 28 November 2022 Accepted - 13 January 2023	Quick Response code
DOI: 10.32677/ijcr.v9i1.3710	

Physical examination revealed an otherwise active child with no major congenital anomalies except for a large abdominal palpable mass in the right lumbar region crossing the midline which is solid in consistency with ill-defined borders. Dilated veins were present over the abdomen (Fig. 1a).

Contrast-enhanced computed tomography abdomen revealed a well-defined retroperitoneal mass lesion of size 7.8 × 5.9 × 7.9 cm, infiltrating the right kidney and adrenal with extension

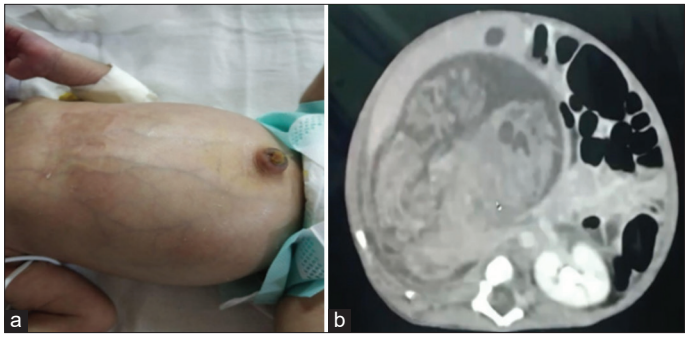

Figure 1: (a) Abdominal mass with dilated veins and (b) computed tomography image of the right renal mass in a 2-day-old neonate

Correspondence to: Teena Rajan, Department of Medical Oncology, Stanley Medical College, Old Jail Road, Old Washermanpet, Chennai - 600 001, Tamil Nadu, India. E-mail: teenarajan97@gmail.com

into the subhepatic space superiorly and inferiorly up to the inferior pole of the right kidney. There was no calcification (Fig. 1b) and the left kidney was normal.

The patient was taken up for right radical nephroureterectomy with a provisional diagnosis of CMN/Wilms tumor. Per-operatively, a 10 × 8 × 8 cm right renal mass was seen infiltrating the right lobe of the liver and attached to the diaphragm (Fig. 2). The tumor was removed in total with anticipated spillage. The post-operative period was uneventful.

Histopathological examination showed triphasic Wilms with a predominant blastemal component and no anaplasia. The tumor weight was 90 g and the size was 9 × 6 × 3.5 cm. The unifocal tumor was extending beyond the pseudocapsule (Fig. 3a). Immunohistochemistry showed WT1 positivity (Fig. 3b). On the basis of this, a final diagnosis of a triphasic Wilms tumor of the right kidney with no anaplasia (COG Stage III, SIOP intermediate risk) was made.

The patient was started on DDA4 (Vincristine, Actinomycin D, Doxorubicin) regimen with 50% dose reduction and completed whole abdominal radiotherapy (10.8 Gy). Further molecular testing for loss of heterozygosity (LOH) of 1p and 16q was sent and was negative for the same (Fig. 3c). She tolerated chemotherapy and radiation well. She is disease free at present and under regular follow-up.

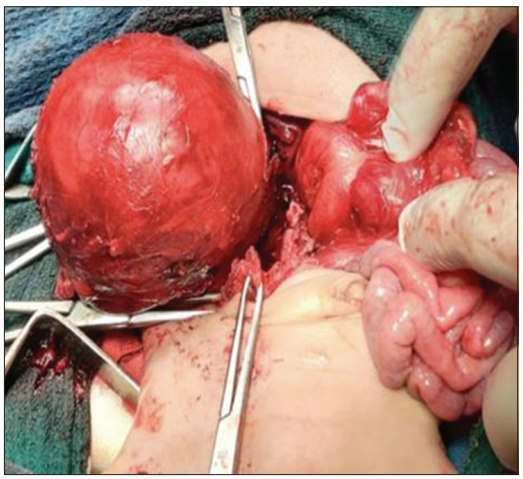

Figure 2: Intraoperative image of the right renal mass in a 2-day-old neonate

DISCUSSION

Neonatal Wilms tumor is a very rare presentation. It is the second most common renal tumor diagnosed in the neonatal age group next to CMN. Wilms tumor develops from the metanephric blastema. It can be associated with other congenital anomalies like WAGR syndrome, Deny Drash syndrome, and Beckwith-Weideman syndrome. Molecular biology lies in the WT1 gene located on chromosome 11p. Other genes involved are WT2, WTX, and the TP53 gene [5]. The most common age of presentation is between 3 and 4 years of age.

Clinically, the child presents with an abdominal mass, gross hematuria, and hypertension [6]. The close differential is neuroblastoma wherein, the child will be sick at presentation. Radiologically, the origin of the lesion and its relation to the nearby structures can be identified which helps in further treatment decisions. There will not be any calcifications within the lesion unlike neuroblastoma [7]. Staging is as per COG guidelines.

Multimodality treatment is necessary for the management of Wilms tumor which is as per SIOP (preop chemo-nephrectomy-adjuvant chemo ± radiotherapy) or NWTS protocol where nephrectomy is the primary treatment followed by adjuvant chemotherapy ± radiotherapy based on the postoperative risk stratification [8]. Most commonly used chemotherapy regimens are EEA4, DDA4, M, and I regimens. The backbone of the regimens is vincristine, actinomycin-D, doxorubicin with or without etoposide, and carboplatin [9]. For infants, the dose is modified by a 50% dose reduction [10]. Adjuvant radiotherapy total dose ranges from 10.8Gy to 19.8Gy depending on the age, stage, and risk stratification [11]. Patients who are positive for LOH of 1p and 16q are associated with inferior survival and may benefit from intensification of adjuvant treatment [12]. Survival rates for non-metastatic Wilms tumor is more than 90% with combined modality treatment. The patient should be closely followed up for recurrences and to monitor the toxicity of the treatment [13].

CONCLUSION

Neonatal Wilms tumor is a very rare presentation. It is indeed a challenge to the oncologists. A dedicated team of pediatric

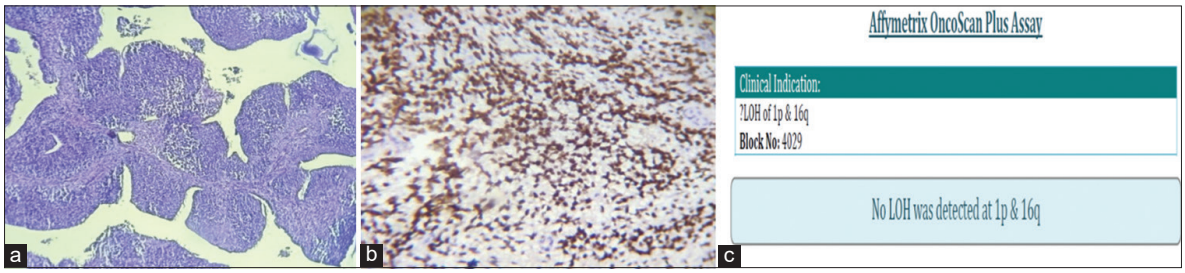

Figure 3: (a) Microscopic appearance of triphasic Wilms tumor; (b) Immunohistochemistry staining positive for wt1; (c) loss of heterozygosity 1p and 16 q testing

surgeons, neonatologists, radiation, and medical oncologists is required for the combined modality treatment. The final results are worth the challenge that is faced.

REFERENCES

1. Glick RD, Hicks MJ, Nuchtern JG, Wesson DE, Olutoye OO, Cass DL. Renal tumors in infants less than 6 months of age. J Pediatr Surg 2004;39:522-5.
2. Zychot E, Apps J, Pritchard-Jones K. Wilms' tumor: Biology, diagnosis and treatment. Transl Pediatr 2014;3:12-24.
3. Spreafico F, Fernandez CV, Brok J, Nakata K, Vujanic G, Geller JI, *et al*. Wilms tumor. Nat Rev Dis Primers 2021;7:75.
4. Ritchey ML, Azizkhan RG, Beckwith JB, Hrabovsky EE, Haase GM. Neonatal Wilms tumor. J Pediatr Surg 1995;30:856-9.
5. Treger TD, Chowdhury T, Pritchard-Jones K, Behjati S. The genetic changes of Wilms tumor. Nat Rev Nephrol 2019;15:240-51.
6. Davidoff AM. Wilms Tumor. Adv Pediatr 2012;59:247-67.
7. Dumba M, Jawad N, McHugh K. Neuroblastoma and nephroblastoma: A radiological review. Cancer Imaging 2015;15:5.
8. Bhatnagar S. Management of Wilms' tumor: NWTS vs SIOP. J Indian Assoc Pediatr Surg 2009;14:6-14.
9. Wang J, Li M, Tang D, Gu W, Mao J, Shu Q. Current treatment for Wilms tumor: COG and SIOP standards. World J Pediatr Surg 2019;2:e000038.
10. Corn BW, Goldwein JW, Evans I, D'Angio GJ. Outcomes in low-risk babies treated with half-dose chemotherapy according to the third national Wilms' tumor study. J Clin Oncol 1992;10:1305-9.
11. Breslow NE, Beckwith JB, Haase GM, Kalapurakal JA, Ritchey ML, Shamberger RC, *et al*. Radiation therapy for favorable histology Wilms tumor: Prevention of flank recurrence did not improve survival on national Wilms tumor studies 3 and 4. Int J Radiat Oncol Biol Phys 2006;65: 203-9.
12. Pan Z, He H, Tang L, Bu Q, Cheng H, Wang A, *et al*. Loss of heterozygosity on chromosome 16q increases relapse risk in Wilms' tumor: A meta-analysis. Oncotarget 2017;8:66467-75.
13. Evans AE, Norkool P, Evans I, Breslow N, D'Angio GJ. Late effects of treatment for Wilms' tumor: A report from the national Wilms' tumor study group. Cancer 1991;67:331-6.

Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Ravel N, Rajan T, Mohankumar K, Vadivelu G, Sathiyamoorthy PN, Ramanan A, Vijayaraghavan P, Joel SM, Chodavarapu D, Konduri M, Pothala V. Wilms tumour in a two day old neonate: A rare and challenging case. Indian J Case Reports. 2023;9(1):20-22.