MAK HILL Publications Research Journal of Medical Sciences

Research Article
doi: 10.36478/makrjms.2025.1.579.582

OPEN ACCESS

Key Words
Malignant rhabdoid tumor, kidney transplant, pediatric donor

Corresponding Author
Amulya Chiliki,
Department of Surgical Oncology, Kims Hospitals, Secunderabad, India

Author Designation
[1]Senior Surgical Oncologist and HOD
[2]Junior Consultant, Surgical Oncology
[3,4]Consultant
[5]Senior Resident
[6-8]Postgraduate Student
[9]Duty Medical Officer

Received: 18 October 2024
Accepted: 21 November 2024
Published: 13 December 2024

Citation: Nagendra Parvataneni, Amulya Chiliki, Diwakar Naidu, Isatish Rao, P. Susmitha, Ishfaq Ahmed, Mahesh Chejarla, G. Reshma Sree and Indrani Biswas, 2025. A Rare and Unusual Occurrence of a Malignant Rhabdoid Tumor in a Pediatric Dual Kidney Transplant Recipient-Challenges in Diagnosis and Management. Res. J. Med. Sci., 19: 579-582, doi: 10.36478/makrjms.2025.1.579.582

A Rare and Unusual Occurrence of a Malignant Rhabdoid Tumor in a Pediatric Dual Kidney Transplant Recipient-Challenges in Diagnosis and Management

[1]Nagendra Parvataneni, [2]Amulya Chiliki, [3]Diwakar Naidu, [4]Isatish Rao, [5]P. Susmitha, [6]Ishfaq Ahmed, [7]Mahesh Chejarla, [8]G. Reshma Sree and [9]Indrani Biswas

[1,2,5-9]Department of Surgical Oncology, Kims Hospitals, Secunderabad, India
[3]Department of Nephrology, Kims, Secunderabad, India
[4]Department of Pathology, kims, Secunderabad, India

ABSTRACT
Renal transplantation is a critical treatment for end-stage renal disease, and pediatric cadaveric dual kidney en bloc transplants provide a valuable option for renal replacement. This case report discusses the rare incidence of a Malignant Rhabdoid Tumor (MRT) in a dual kidney transplant recipient, emphasizing the challenges in diagnosis and management. A 57-year-old female, with a history of hypertension and chronic kidney disease, received a pediatric cadaveric dual kidney en bloc transplant. Eleven months post-transplant, she developed severe abdominal pain. Imaging suggested a hypodense lesion in the transplanted kidney. Differential diagnosis considered post-transplant lymphoproliferative disorder and Wilms tumor. Following a graft nephrectomy due to complications, histopathology confirmed an MRT. Management included extensive surgery, immunosuppressive adjustment and adjuvant therapy. Immunol is to chemistry indicated loss of INI-1 expression in the tumor, crucial for the diagnosis. MRT in transplanted kidneys is an unusual but serious complication post-transplant. This case underscores the necessity for vigilant follow-up, comprehensive diagnostic strategies and individualized treatment plans to manage such rare occurrences effectively.

INTRODUCTION

Renal transplantation remains a cornerstone in the management of end-stage renal disease (ESRD). The use of paediatric cadaveric dual kidneys en bloc offers a viable option for patients requiring a renal replacement therapy. However, such procedures can lead to unique postoperative complications and management challenges. This case report details a patient who had a Rhabdoid tumor in the transplanted kidney, following a dual kidney en bloc transplant.

Case Report: A 57-year-old female with a history of hypertension and chronic kidney disease, vaccinated for Pneumococcus, maintained on dialysis for 6 years, underwent paediatric cadaveric dual kidney en bloc transplantation.

Surgical Procedure: The dual kidneys were placed en bloc in the iliopsoas and true pelvis. Anastomoses included the donor abdominal aorta to the external iliac artery (end-to-side) and the donor IVC to the external iliac vein (end-to-side). Ureters were joined by pantaloons technique. Double J stent placed. The patient exhibited good diereses intra operatively.

Postoperative Course: The patient received Thymoglobulin induction and was placed on a triple-drug immunosuppressive regimen (MMF, Tacrolimus, Wysolone). Postoperatively, she experienced delayed graft function with reduced urine output, necessitating two hemodialysis sessions. By postoperative day 5, graft function improved, with serum creatinine levels decreasing to 3.3 mg/dL at discharge.

Follow-Up and Complications: Eleven months post-transplant, the patient presented with severe diffuse abdominal pain. CT imaging revealed a large, well-defined, hypodense lesion involving the interpolar region and hilum of the transplanted kidney, with indistinct fat planes involving the sigmoid colon. Differential diagnoses included post-transplant lymphoproliferative disorder (PTLD) or Wilms tumor.
Surgical Intervention: Upon Laparotomy, a graft nephrectomy was performed due to one of the kidneys rupturing and harbouring a venous thrombosis. Postoperatively, the patient's urine output improved, and she stabilized. Postoperative histopathology was confirmatory of a rhabdoid tumor of the kidney. One month later, a PET scan identified a large lobulated mass in the right para iliac region extending into the pelvis, with indistinct fat planes with uterus and sigmoid colon, with the presence of enlarged right common and external iliac lymph nodes. An exploratory laparotomy with mass excision, right pelvic lymphadenectomy (RPLND) and pelvic nodal dissection was conducted. The mass was densely adherent to the uterus, presacral fascia, mesorectal fascia, hypogastric nerve plexus and transplant structures. The rectal deposit was excised with a primary closure, hysterectomy, bilateral salpingo-oophorectomy (BSO), and a defunctioning stoma was created. The transplant kidney, vessels, ureter and anastomosis were preserved.

Postoperative Course: The patient developed atrial fibrillation (AF) in the post operative period which was managed appropriately. Immunosuppressants and antihypertensives were restarted. Histopathology-Microscopically, renal parenchyma showed infantile glamorously. Occasional foci showed infiltrate of atypical cells arranged in sheet like pattern. The cells have irregular nuclei, few having vesicular chromatin, conspicuous nucleoli with scant to moderate rim of eosinophilic cytoplasm. Few cells show rhabdoid morphology. Immunohistochemistry-CD3, CD20, CD30, LCA, Synaptophysin, SS18-SSX, S100, CD34, CD117, Myogenin are negative in the neoplastic cells. INI-1 expression is lost in the neoplastic cells. Desmin shows paranuclear dot like staining. PAN CK shows staining in rare cells. Ki 67 index is high, about 55%.

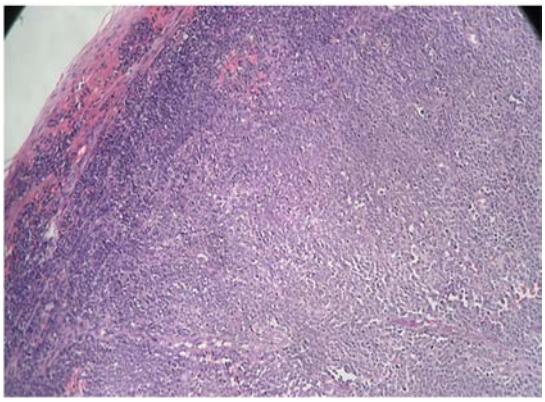

Fig. 1: Low Magnification Photomicrograph Showing the Architectural Pattern of the Tumor with a Diffuse Growth Pattern

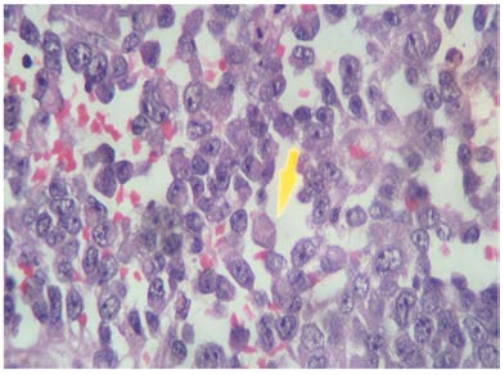

Fig. 2: High Magnification Photomicrograph of the Tumor, Highlighting the Large, Eccentric Nuclei with Prominent Nucleoli and Cytoplasmic Eosinophilic Inclusions (Yellow Arrow Pointing to a Rhabdoid Cell)

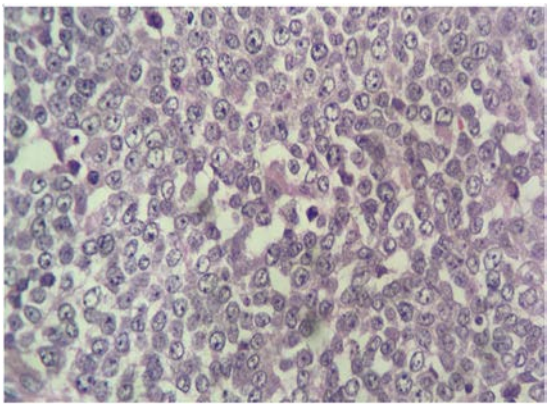

Fig. 3: Close-Up View of the Malignant Rhabdoid Cells Showing Pleomorphic, Round to Oval Nuclei, with Distinct Borders and Coarse Chromatin, a Hallmark Feature of Rhabdoid Tumors

Primary tumors in the kidney allograft recipients have often been reported since the advent of potent immunosuppression. The most common malignant tumors of organ transplant recipients are post-transplant lymphoproliferative diseases and skin cancer[19]. To our knowledge, this is the second case of MRT being reported in a kidney transplanted into an adult. MRT of the kidney usually affects children <2 years of age[1]. It comprises 1.8% of all pediatric renal tumors in the National Wilms' Tumor Study[2] Adult renal MRT is even less common and only six cases have been reported[1,3]. Age at diagnosis has ranged from 32-60 years with no predominant gender. The postdiagnosis survival time was usually only a few months. Criteria essential for pathologic diagnosis of MRT include characteristic histology showing rhabdoid features (as described above) and immunohistochemistry indicating loss of INI-1 protein nuclear expression. Although not essential for diagnosis, negative myogenic markers and electron microscopic confirmation of cytoplasmic intermediate filaments in swirls or whorls provide additional support for this diagnosis. Also, loss of INI-1 is helpful to differentiate MRT from other primary renal neoplasms[4-7] with rhabdoid differentiation including clear cell, transitional cell, papillary, chromophobe and collecting duct carcinomas[8-10]. Rhabdoid tumor in a transplanted kidney can present with a variety of symptoms, including abdominal pain, palpable mass, or signs of graft dysfunction. As seen in this case, the patient presented with severe diffuse abdominal pain and a large hypodense lesion in the transplanted kidney area. Differential diagnoses for a mass in a transplanted kidney include post-transplant lymphoproliferative disorder (PTLD), Wilms tumor and other renal malignancies. PTLD is particularly relevant in the post-transplant setting due to its association with Epstein-Barr virus (EBV) and immunosuppressive therapy. Distinguishing between Malignant Rhabdoid tumor and PTLD can be challenging. PTLD typically presents with a more diffuse pattern of involvement, whereas MRT tends to form localized, well-defined masses. A definitive diagnosis of MRT requires histological examination of the tumor. Immunohisto chemical staining and molecular studies are essential for accurate classification and to differentiate MRT from other neoplasms. In the present case, we observed loss of INI1 expression in the tumour. It was also observed that the normal cells in the kidney of the patient expressed INI-1. Therefore, loss of INI1 was specific to the tumour cells. Heterozygous abnormality of the INI1 gene has been reported in some children with MRTs [11-16]. It has been reported that some adult MRT types coexist with other types of RCCs. In these cases, MRT is hypothesized to develop from RCC. Therefore, it is recommended to examine INI1 expression in MRTs combined with other RCC types. Other malignancies, including rhabdomyosarcoma, myoepithelioma in soft tissue, epithelioid angiosarcoma and epithelioid malignant peripheral nerve sheath tumor (MPNST), can be distinguished from MRT by their specific markers and INI1 expression. Loss of INI1 is also helpful to differentiate MRT from other primary renal neoplasms with rhabdoid differentiation, including clear cell renal carcinoma, chromophobe cell renal carcinoma and other malignance in the urinary system[17,18]. The primary treatment for MRT is surgical resection. In this case, extensive surgery with exploratory laparotomy, mass excision and lymphadenectomy was required due to the mass's adherence to surrounding structures. Adjuvant therapy typically includes chemotherapy and in some cases, radiation therapy. The specifics of adjuvant therapy in the context of a renal transplant must be carefully balanced with the need to maintain graft function and manage immunosuppression. Adjusting immunosuppressive therapy is vital in managing post-transplant malignancies. A multidisciplinary team, including oncoloists and transplant nephrologists, must coordinate to optimize treatment while minimizing the risk of graft rejection.

Prognosis: The prognosis of MRT in the transplant setting depends on various factors, including the tumor's stage, response to treatment and the patient's overall health. The rarity of RMS in transplanted kidneys necessitates individualized treatment plans and close monitoring.

Follow-Up: Long-term follow-up is essential for monitoring for recurrence of the malignancy and ensuring ongoing graft function. Regular imaging

studies and clinical evaluations are necessary to detect any residual or recurrent disease.

CONCLUSION

MRT arising in a transplanted kidney represents a rare but significant complication in post-transplant care. This case underscores the importance of considering a broad differential diagnosis when evaluating masses in transplanted organs. Effective management requires a multidisciplinary approach, balancing the need for oncologic treatment with the preservation of graft function and overall patient health. Continued research and case reporting will enhance understanding and management of such rare occurrences, improving outcomes for future patients.

REFERENCES

1. Peng, H.Q., A.E. Stanek, S. Teichberg, B. Shepard and E. Kahn, 2003. Malignant Rhabdoid Tumor of the Kidney in an Adult: A Case Report and Review of the Literature. Arch. Pathol. And Lab. Med., 127: 371-373.
2. ZHAO, G., R. NA, Y. YANG and R. HAN, 2013. Pure malignant rhabdoid tumor of the left kidney in an adult: A case report and review of the literature. Oncol. Lett., 5: 1481-1484.
3. Kang, Y.N., S.P. Kim and B.K. Jang, 2013. A Primary Malignant Rhabdoid Tumor in Adult Liver. Korean J. Pathol., 47: 468-488.
4. Versteege, I., N. Sévenet, J. Lange, M.F. Rousseau-Merck and P. Ambros *et al.,* 1998. Truncating mutations of hSNF5/INI1 in aggressive paediatric cancer. Nature, 394:203-206.
5. Ogino, S., J.Y. Ro and R.W. Redline, 2000. Malignant Rhabdoid Tumor: A Phenotype? An Entity?-A Controversy Revisited. Adv. Anatomic Pathol., 7: 181-190.
6. Perry, A., C.E. Fuller, A.R. Judkins, L.P. Dehner and J.A. Biegel, 2005. INI1 expression is retained in composite rhabdoid tumors, including rhabdoid meningiomas. Mod. Pathol., 18: 951-958.
7. Kumar, S., D. Kumar and D.F. Cowan, 1992. Transitional Cell Carcinoma With Rhabdoid Features. The Am. J. Surg. Pathol., 16: 515-521.
8. Weeks, D.A., J.B. Beckwith, G.W. Mierau and C.W. Zuppan, 1991. Renal Neoplasms Mimicking Rhabdoid Tumor of Kidney. The Am. J. Surg. Pathol., 15: 1042-1054.
9. Yusuf, Y., A.H. Belmonte and V. Tchertkoff, 1996. Fine Needle Aspiration Cytology of a Recurrent Malignant Tumor of the Kidney with Rhabdoid Features in an Adult. Acta Cytologica, 40: 1313-1316.
10. Roberts, K.E., D. Hamele-Bena, A. Saqi, C.A. Stein and R.P. Cole, 2003. Pulmonary tumor embolism: A review of the literature. The Am. J. Med., 115:228-232.
11. Kane, R.D., H.K. Hawkins, J.A. Miller and P.S. Noce, 1975. PULMONARY TUMOR EMBOLI. Cancer, 36: 1473-1482.
12. Winterbauer, R.H., I.B. Elfenbein and W.C. Ball, 1968. Incidence and clinical significance of tumor embolization to the lungs. The Am. J. Med., 45: 271-290.
13. Keenan, N.G., A.G. Nicholson and P.J. Oldershaw, 2008. Fatal acute pulmonary hypertension caused by pulmonary tumour thrombotic microangiopathy. Int. J. Cardiol., 124: 11-13.
14. Sakashita, N., C. Yokose, K. Fujii, M. Matsumoto, K. Ohnishi and M. Takeya, 2007. Pulmonary tumor thrombotic microangiopathy resulting from metastatic signet ring cell carcinoma of the stomach. Pathol. Int., 57:383-387.
15. Biegel J.A, J.Y. Zhou, L.B. Rorke, C. Stenstrom, L.M. Wainwright and B. Fogelgren. 1999. Germ-line and acquired mutations of INI1 in atypical teratoid and rhabdoid tumors. Canc. Res., 59: 74-79.
16. Przybycin, C.G., J.K. McKenney, J.P. Reynolds, S. Campbell, M. Zhou, M.T. Karafa and C. Magi-Galluzzi, 2014. Rhabdoid Differentiation Is Associated With Aggressive Behavior in Renal Cell Carcinoma. Am. J. Surg. Pathol., 38: 1260-1265.
17. Shannon, B.A. and R.J. Cohen, 2003. Rhabdoid differentiation of chromophobe renal cell carcinoma. Pathology, 35: 228-230.
18. Saleeb, R., H. Faragalla, G. Yousef, R. Stewart and C. Streutker, 2016. Malignancies in a renal transplant population: The St. Michael's Hospital experience. Urol. Ann., 8: 163-167.