Non-syndromic first case of pediatric rhabdomyosarcoma originating from the umbilical left medial ligament: A case report

MELTEM KABA[1], ÇETIN ALI KARADAĞ[1], SERKAN ODABAŞI[1] and DILDAR BAHAR GENÇ[2]

Departments of [1]Pediatric Surgery and [2]Pediatric Oncology, Sisli Hamidiye Etfal Training and Research Hospital, University of Health Sciences, Sarıyer, 34453 İstanbul, Turkey

Received July 3, 2023; Accepted November 8, 2023

DOI: 10.3892/ol.2023.14171

Abstract. Rhabdomyosarcoma (RMS) is the most common soft-tissue sarcoma in children. The present study reports the case of a 2-year-old female who presented with abdominal pain and a palpable abdominal mass. Radiological investigations failed to reveal the tissue origin of the mass and a tru-cut biopsy confirmed the diagnosis of embryonal RMS. Surgical excision was performed after neo-adjuvant chemotherapy. The pelvic end of the mass was observed to continue with the left medial umbilical ligament. The patient's postoperative course was uneventful, and follow-up imaging showed no evidence of recurrence. The present case report is the first non-syndromic case with left umbilical medial ligament-originated RMS.

Introduction

Rhabdomyosarcoma (RMS), the most common soft-tissue sarcoma in children and adolescents, originates from mesenchymal cells and can be found anywhere in the body (1). RMS is most commonly located in the head and neck region and genitourinary system, while it is rarely observed in the pelvis (2). RMS originating from urachal remnants has been reported in the pediatric age group. Almost all cases reported in the literature consist of children with Costello Syndrome (CS) who tend to have malignant tumors (3). The medial umbilical ligaments are paired structures related to the umbilical arteries found either side of the median umbilical ligament. The median and medial umbilical ligaments form a peritoneal depression on each side of the urinary bladder referred to as the supravesical fossae (4). RMS originating from the umbilical remnants is an exceedingly rare entity, with limited reported cases in the literature. To the best of our knowledge, there is no case of medial ligament-originated RMS and also non-syndromic in the literature review. The challenges of identifying the tissue origin of the mass, especially in large tumors, highlight the importance of a thorough exploration by the surgeon during the procedure. In the present case, the surgeon played a crucial role in determining the attachment of the mass to tubular structures and therefore guiding the appropriate intervention.

Case report

The current study reports the case of a previously healthy 2-year-old girl who was the first child of healthy and non-consanguineous parents. The prenatal follow-ups and birth history were uneventful. The growth and development examinations were appropriate for the patient's age. The patient was admitted to the Emergency Department of Sisli Hamidiye Etfal Training and Research Hospital (Istanbul, Turkey) in March 2020 with ongoing abdominal pain for a week and abdominal distension, which were considered to be due to weight gain for a while. The patient was transferred to the Department of Pediatric Surgery, Sisli Hamidiye Etfal Training and Research Hospital, after an 11-cm mass filling the entire pelvis was identified on Doppler ultrasonography. The abdominal magnetic resonance imaging (MRI) (Fig. 1) revealed that the 11x8-cm solid mass with smooth borders and possibly encapsulated was compressing the sigmoid colon. The mass could not be separated from the bladder and had high contrast, and it was reported that it could be either an ovarian solid tumor or bladder-originated RMS. It was also stated that the bilateral pelvic ectasia detected in the patient was probably due to the compression of the mass. Tru-cut biopsy was taken from the mass with an 18-gauge needle under ultrasonography guidance. The pathological examination reported a small round-cell malignant tumor. After immunohistochemical staining performed by the Pathology Department, the patient was diagnosed with embryonal-type RMS pathologically. As a result of the other examinations performed by a pediatric oncologist, it was observed that there was no metastasis. Since the primary mass was unsuitable for complete resection, the patient was accepted as Intergroup RMS Studies (IRS) Group III, and neoadjuvant chemotherapy was initiated according to the COG D9803 protocol (5). The patient was re-evaluated

Correspondence to: Dr Meltem Kaba, Department of Pediatric Surgery, Sisli Hamidiye Etfal Training and Research Hospital, University of Health Sciences, 64 Bahçeköy Street, Sarıyer, 34453 Istanbul, Turkey
E-mail: meltemkaba@windowslive.com

Abbreviations: RMS, rhabdomyosarcoma; CS, Costello Syndrome; MRI, magnetic resonance imaging

Key words: RMS, CS, childhood, urachus, left medial ligament

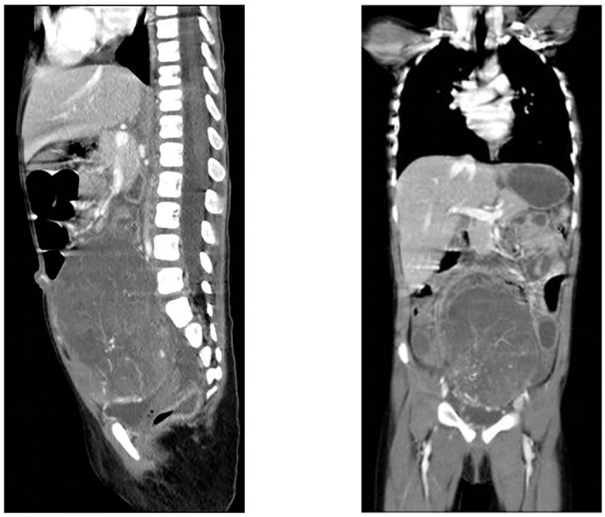

Figure 1. Magnetic resonance imaging of the patient before chemotherapy.

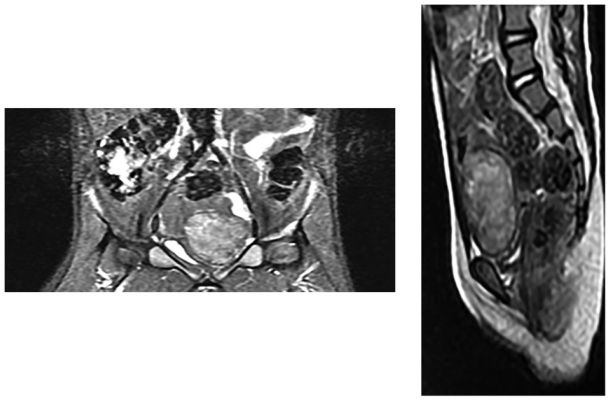

Figure 2. Magnetic resonance imaging after chemotherapy. There was a heterogeneous, thick-walled, heterogeneous cystic mass in the pelvis, ~45 mm in diameter, which compressed the bladder.

after 4 sessions of neoadjuvant chemotherapy with VAC (vincristine, Actinomycin D and Cyclophosphamide). MRI images after the chemotherapy are presented in Fig. 2.

The patient underwent a diagnostic cystoscopy before surgery. It was observed that the bladder bulged inward from the left lateral wall, and the bladder mucosa was normal. After cystoscopy, the mass was freed from peritoneal adhesions on the left lateral abdominal wall and the posterior wall of the bladder by laparoscopic surgery. The pelvic end of the mass was observed to continue with the left medial umbilical ligament. The left medial umbilical ligament was ligated and cut, and the mass was excised with a mini-pfannenstiel incision (Fig. 3). The oral intake of the patient was initiated on the first day after surgery. The patient's wound remained clean, and was transferred to the oncology service two days after the surgery. The pathology result of the mass was reported as embryonal-type RMS. No residual tumor was observed in the postoperative imaging. A total of four sessions of the VAC chemotherapy protocol were continued after surgery (0.05 mg/kg vincristine, 0.045 mg/kg Actinomycin D, 73 mg/kg Cyclophosphamide). Moreover, 50.4 Gy of radiotherapy was applied to the tumor site and

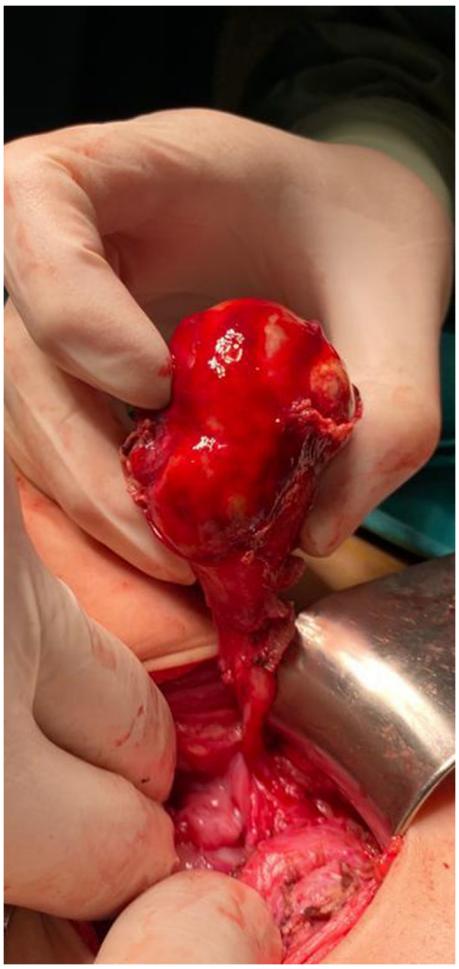

Figure 3. Surgical image of the mass originating from the left medial ligament.

18 Gy to the entire abdomen. After completing her treatment, the patient had been followed up for 2 years and 3 months without any disease.

Discussion

RMS is the most common soft tissue sarcoma of childhood. The incidence is 4 to 7 cases per 1 million children younger than 15 years, and the peaks occur between the ages of 2 and 5 years (3). Similar to other soft-tissue sarcomas, RMS originates most commonly from the extremities, followed by the trunk wall, retroperitoneum, genitourinary tract, head, and neck. Primary pelvic RMS is extremely rare, included within the so-called 'Other sites' as abdominal or thoracic locations (6). Primary pelvic RMS, mostly observed in paediatric patient groups (7), poses a great challenge for clinicians. Tumors in pelvic sites often grow and spread locally without symptoms, and the exact site of origin is challenging to determine at the time of diagnosis.

The umbilical artery is located within the umbilical cord and carries deoxygenated blood from the fetus to the mother. After the birth, the distal portion obliterates as the medial umbilical ligament within a fold of the peritoneum. It lies lateral to the median umbilical ligament (urachus). The persistent proximal portion of the umbilical artery runs along the side wall of the

pelvis as the superior vesical artery (8). The varied morphology of the umbilical ring and its surrounding structures are still being investigated to deepen understanding of this complicated anatomical region (9,10). Further studies may also shed light on this unique site selection of RMS in these patients.

In the general population, umbilical ligament malignant tumors are extremely rare, usually observed in adult males, accounting for 0.01% of all tumors (11). On the other hand, umbilical ligament RMS is mostly reported in paediatric patients with CS. In a literature review published in 2017, the total number of cases of RMS originating from the umbilical ligaments in paediatric patients was only 17. A total of 15 cases originated from the urachus, and two were from the medial umbilical ligaments. Both of these cases were patients with CS (12,13). Previously, only 12 children with no CS with urachus-originated RMS have been reported in the literature. A case of RMS originating from the medial umbilical ligament without CS has not been encountered in the literature; therefore, the present case report was the first reported in this respect.

CS is a rare neurodevelopmental disorder caused by germline mutations in *HRAS*. Typical features include a distinctive facial appearance, growth retardation, intellectual disability, ectodermal, cardiac, musculoskeletal abnormalities and cancer susceptibility. A total of ~10% of children with CS develops cancer. A total of ~60% of these cancers are embryonal RMS and RMSs, specifically originating from umbilical remnants in children with CS. CS had been excluded from the present case report, and no other genetic abnormalities were investigated.

The study presenting the largest series of pediatric RMS from the urachal origin with 8 cases proved that urachal localization had a worse prognosis (14). It has been argued that RMS originating from the umbilical ligaments worsens the prognosis by allowing metastasis at the time of diagnosis since it presents with large asymptomatic masses in the preperitoneal area. Although the present case report presented a mass reaching 11 cm in diameter, there was no distant metastasis and local spread, and she remained disease and complaint-free for 3 years after treatment. The other two cases in the literature originating from the medial umbilical ligament were also disease-free during their four-year follow-up. Although the number of cases is too small to be argued about the prognosis, it may be necessary to address the prognosis difference between the two localizations despite the anatomical, embryogenetic, and histological similarities. In order to evaluate the prognosis of this localization, it is crucial to include rare disease presentations in the literature. As the number of patients and data increases, identifying these cases clearly instead of classifying them within the large so-called 'others' group may help the treatment management of these patients.

The organ or tissues from which the mass originates may not be clear in large tumors. In the present case report, the surgeon is the key person to determine which tubular structure the mass was attached to after exploring and identifying other pelvic tubular organs. This situation can be decided with the basic surgical principle to completely or partially excise the organ originating from the mass considered malignancy, as a result, to be prepared for very rare situations that may be encountered for the first time in the operating room. Therefore, it is vital that rare disease presentations that cannot be serialized should be included in the literature and that surgeons' attention should be drawn to the subject.

Acknowledgements

Not applicable.

Funding

No funding was received.

Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Authors' contributions

MK conceived and designed the study, analyzed and interpreted the results, and prepared the draft of the manuscript. CAK obtained medical images and analysed the results. SO performed data collection. DBG advised on patient treatment or analyzed patient data. All authors reviewed the results, and read and approved the final version of the manuscript. MK, CAK, SO and DBG confirm the authenticity of all the raw data.

Ethics approval and consent to participate

Written informed consent was obtained from the legal guardians of the patient for participation.

Patient consent for publication

Written informed consent was obtained from the legal guardians of the patient for publication of the case information and associated images.

Competing interests

The authors declare that they have no competing interests.

References

1. Howlader N Noone AM, Krapcho M, Miller D, Bishop K, Kosary CL, Yu M, Ruhl J, Tatlovich Z, Mariotto A, Lewis DR, *et al*: SEER Cancer Statistics Review, 1975-2014. National Cancer Institute, Bethesda, MD, USA. https://seer.cancer.gov/csr/1975_2014/ based on November 2016 SEER data submission, posted to the SEER website, April 2017.
2. Shern JF, Yohe ME and Khan J: Pediatric Rhabdomyosarcoma. Crit Rev Oncog 20: 227-243, 2015.
3. Gripp KW, Scot CI Jr, Nicholson L, McDonald-McGinn DM, Ozeran JD, Jones MC, Lin AE and Zackai EH: Five additional Costello syndrome pa=tients with rhabdomyosarcoma: Proposal for a tumor screening protocol. Am J Med Genet 108: 80-87, 2002.
4. Moore KL, Dalley AF and Agur AMR: Clinically Oriented Anatomy. 7th edition. Lippincott Williams & Wilkins, Philadelphia, PA, USA, 2014.
5. Arndt CA, Stoner JA, Hawkins DS, Rodeberg DA, Hayes-Jordan AA, Paidas CN, Parham DM, Teot LA, Wharam MD, Breneman JC, *et al*: Vincristine, actinomycin, and cyclophosphamide compared with vincristine, actinomycin, and cyclophosphamide alternating with vincristine, topotecan, and cyclophosphamide for intermediate-risk rhabdomyosarcoma: Children's oncology group study D9803. J Clin Oncol 27: 5182-5188, 2009.
6. Rodary C, Flamant F and Donaldson SS: An attempt to use a common staging system in rhabdomyosarcoma: A report of an international workshop initiated by the International Society of Pediatric Oncology (SIOP). Med Pediatr Oncol 17: 210-215, 1989.

7. Hao Z and Yang S: Embryonal rhabdomyosarcoma within abdomen and pelvis in an adult. Int J Immunopathol Pharmacol 32: 2058738418806728, 2018.
8. Shiffman MA: Pediatric Umbilical Reconstruction: Principles and Techniques. August 2017. Doi: 10.1007/978-3-319-43890-0ISBN: 978-3-319-43888-7.
9. Oh CS, Won HS, Kwon CH and Chung IH: Morphologic variations of the umbilical ring, umbilical ligaments and ligamentum teres hepatis. Yonsei Med J 49: 1004-1007, 2008.
10. Kim JH, Hayashi S, Jin ZW, Murakami G and Rodríguez-Vázquez JF: Umbilical cord vessels other than the umbilical arteries and vein: A histological study of midterm human fetuses. Anat Cell Biol 55: 467-474, 2022.
11. Fernández EM, Siverio NH, Almaraz RL, Viota LM, Luis JR and Flores LD: Radical surgery and IVA-chemotherapeutic regimen to treat embryonal rhabdomyosarcoma of the urachus: Case report. Pediatr Hematol Oncol 24: 543-550, 2007.
12. Bisogno G, Murgia A, Mammi I, Strafella MS and Carli M: Rhabdomyosarcoma in a patient with cardio-facio-cutaneous syndrome. J Pediatr Hematol Oncol 21: 424-427, 1999.
13. Sánchez-Montenegro C, Vilanova-Sánchez A, Barrena-Delfa S, Tenorio J, Santos-Simarro F, García-Miñaur S, Lapunzina P and Martínez-Martínez L: Costello syndrome and umbilical ligament rhabdomyosarcoma in two pediatric patients: Case reports and review of the literature. Case Rep Genet 2017: 1587610, 2017.
14. Cheikhelard A, Irtan S, Orbach D, Minard-Colin V, Rod J, Martelli H and Sarnacki S: Urachal rhabdomyosarcoma in childhood: A rare entity with a poor outcome. J Pediatr Surg 50: 1329-1333, 2015.