Surgical and Experimental Pathology

CASE REPORT

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Urachal mucinous cystic tumor of low malignant potential: a report of a rare case with literature review

Mthabisi Talent George Moyo[1], Fikret Dirilenoğlu[2*] and Yazgı Köy[3]

Abstract

Neoplasms originating from the urachus are exceptionally uncommon, comprising only 0.17% of all bladder neoplasms. Among these, the mucinous cystic tumor of low malignant potential (MCTLMP) is particularly rare, with only 25 documented cases in medical literature. Despite their rarity, it is essential to identify MCTLMPs given the possibility of severe complications. Fortunately, surgical removal offers promising cure rates. In this report, A 40-year-old female patient presented to the hospital with abdominal pain. Subsequently, a CT scan showed a 9-cm cystic mass pressing the bladder. The lesion was surgically excised and histopathological examination was performed. Grossly, the specimen consisted of a cyst with a smooth external surface and mucinous content. Microscopically, the cyst was lined by columnar mucinous epithelial cells that included basally-located hyperchromatic nuclei and scattered Goblet cells consistent with an intestinal phenotype. The epithelial cells show mild atypia with focal papillary formations and flattening due to compression. Immunohistochemically, the epithelial tumor cells expressed cytokeratin 20, CDX2, beta-catenin (membranous); and were negative for cytokeratin 7. The location, histopathological, and immunohistochemical findings were consistent with a diagnosis of MCTLMP. In this report, we present an instance of urachal MCTLMP, which represents less than 30 cases documented in the medical literature. This case study marks the importance of early identification of MCTLMP due to their potential for severe complications, despite the tumor's low malignant potential and to exclude more aggressive tumors with areas of intraepithelial or invasive carcinoma.

Keywords Urachus, Mucinous cystic tumor of low malignant potential, Bladder, Pathology

Background

Urachal neoplasms are thought to arise from neoplastic transformation of remnant urachal tissue left from incomplete regression of the urachus in foetal development (Brennan et al. 2019). The urachus is an embryologic remnant of the allantois, which connects the anterior dome of the bladder to the umbilicus and rarely persists after early infancy (Schmeusser et al. 2022). In the event of incomplete atresia, the urachus has the potential to transform into numerous pathologies including fistula, diverticulum, or tumor (Wu et al. 2017). Among these pathologies of urachal remnants, urachal

*Correspondence:
Fikret Dirilenoğlu
fikret.dirilenoglu@neu.edu.tr
[1]Department of Biomedical Engineering, Faculty of Engineering, Near East University, Nicosia, Cyprus
[2]Department of Pathology, Faculty of Medicine, Near East University, Nicosia, Cyprus
[3]Department of Pathology, Batman Training and Research Hospital, Batman, Turkey

neoplasms are extremely uncommon and are of significant interest due to their potentially aggressive and invasive behavior. Urachal mucinous tumors are exceedingly rare, representing 0.17% of all bladder cancers (Schmeusser et al. 2022). Their behaviour can range from relatively benign to malignant that can spread distantly or throughout the peritoneum as pseudomyxoma peritonei or peritoneal carcinomatosis (Wang et al. 2016).

Most urachal neoplasms are epithelial (glandular) neoplasms, typically with an intestinal phenotype (Bruins et al. 2012). The spectrum of cystic urachal mucinous neoplasms, including mucinous cystadenoma, MCTLMP, and mucinous cystadenocarcinoma, is similar to the morphologic spectrum of appendiceal and ovarian intestinal-type mucinous neoplasms (Brennan et al. 2019). Consequently, the absence of a known primary glandular neoplasm at another anatomical site has been put forward as a criterion for pathologic diagnosis of a urachal mucinous neoplasm (Brennan et al. 2019).

According to our literature survey, less than 70 urachal neoplasms have been reported in English literature, among which include just 25 prior known cases of MCTLMP first reported in 2006 (Brennan et al. 2019). Here, we presented a case of a MCTLMP in a 40-year-old female with clinical, radiological, and detailed histopathological features. Although rare, MCTLMPs are important to identify due to their potential devastating complications and high rates of recovery after surgical removal (Schmeusser et al. 2022).

Case presentation

A 40-year-old female patient presented to the hospital with abdominal pain. Subsequently, an abdominal CT scan showed a cystic mass of 9×8.5×8 cm in size in the pelvic region, containing septations and applying pressure to the bladder as illustrated in Fig. 1a. The lesion was surgically excised and sent to the pathology lab. Grossly, the specimen consisted of a cyst with a smooth external surface and mucinous content. Microscopically, the cyst was lined by a mucinous epithelium that included basally-located hyperchromatic nuclei and scattered Goblet cells consistent with an intestinal phenotype as shown in Fig. 1b. The epithelial cells show mild atypia with focal papillary formations and flattening due to compression as seen in Fig. 1c. Immunohistochemically, the epithelium expressed beta-catenin (membranous), cytokeratin 20, CDX2; and was negative for cytokeratin 7 as illustrated in Fig. 1c **and** Fig. 1d. The location, histopathological, and immunohistochemical findings were consistent with a diagnosis of "urachal mucinous cystic tumor with low malignant potential." No recurrence or metastasis occurred in 32 months of follow-up.

Discussion and conclusion

MCTLMPs originating from the urachus are exceedingly rare, representing only a minute fraction of all bladder neoplasms (Schmeusser et al. 2022). In this report, we present a case of a 40-year-old female patient who presented with abdominal pain and was subsequently diagnosed with MCTLMP. The limited number of reported cases, with only 26 instances documented in medical literature, underscores the need for heightened awareness and understanding of this unique entity.

Histologically, MCTLMPs exhibit a characteristic cystic architecture with mucinous epithelial lining. Additional features, such as focal papillary formations and compression-induced flattening, provide valuable support for the diagnosis. The immunohistochemical (IHC) profile of the tumor, including positive expression of beta-catenin (membranous), cytokeratin 20, and CDX2, while being negative for cytokeratin 7, aligns with previously reported cases, confirming the mucinous origin and helping to differentiate it from other primary glandular neoplasms in different anatomical sites (Schmeusser et al. 2022). Extensive sampling with careful histopathological examination is essential to exclude areas of intraepithelial or invasive carcinoma (Brennan et al. 2019).

An intriguing aspect of diagnosing MCTLMP and other urachal neoplasms lies in its heavy reliance on histological features, as illustrated in our case. Mucinous cystic tumors are classified similarly to ovarian mucinous cystic tumors, with MCTLMP defined as a "cystic tumor with areas of epithelial proliferation, including papillary formation and low-grade atypia (Agnihotri et al. 2020). The linings of MCTLMPs show higher proliferation rates compared to cystadenomas, ranging from flat to tufted and displaying pseudopapillary to tubulovillous features (Schmeusser et al. 2022).

Immunohistochemical analysis has not been suggested initially or in confirmatory examinations, as it seems to offer limited diagnostic value for urachal neoplasms (Wang and Sule 2018). A study investigating the IHC expression of urachal carcinomas for improved neoplasm delineation revealed an overlapping immunoprofile, providing little clinical benefit (Paner et al. 2016). However, in cases presenting as distantly metastasized cystadenocarcinomas, IHC might be necessary to ascertain tissue origin. In urachal mucinous cystic tumors, positive staining for cytokeratin 20, CDX2, and cytokeratin 7 is observed in 100%, 80%, and 30% of cases, respectively, with negative nuclear β-catenin, estrogen receptor, and progesterone receptor staining (Wang and Sule 2018).

Considering the potential for complications, growth, and increased malignancy, surgical management of mucinous urachal neoplasms is imperative. MCTLMP should be surgically excised through partial cystectomy, followed by regular imaging and cystoscopy for recurrence

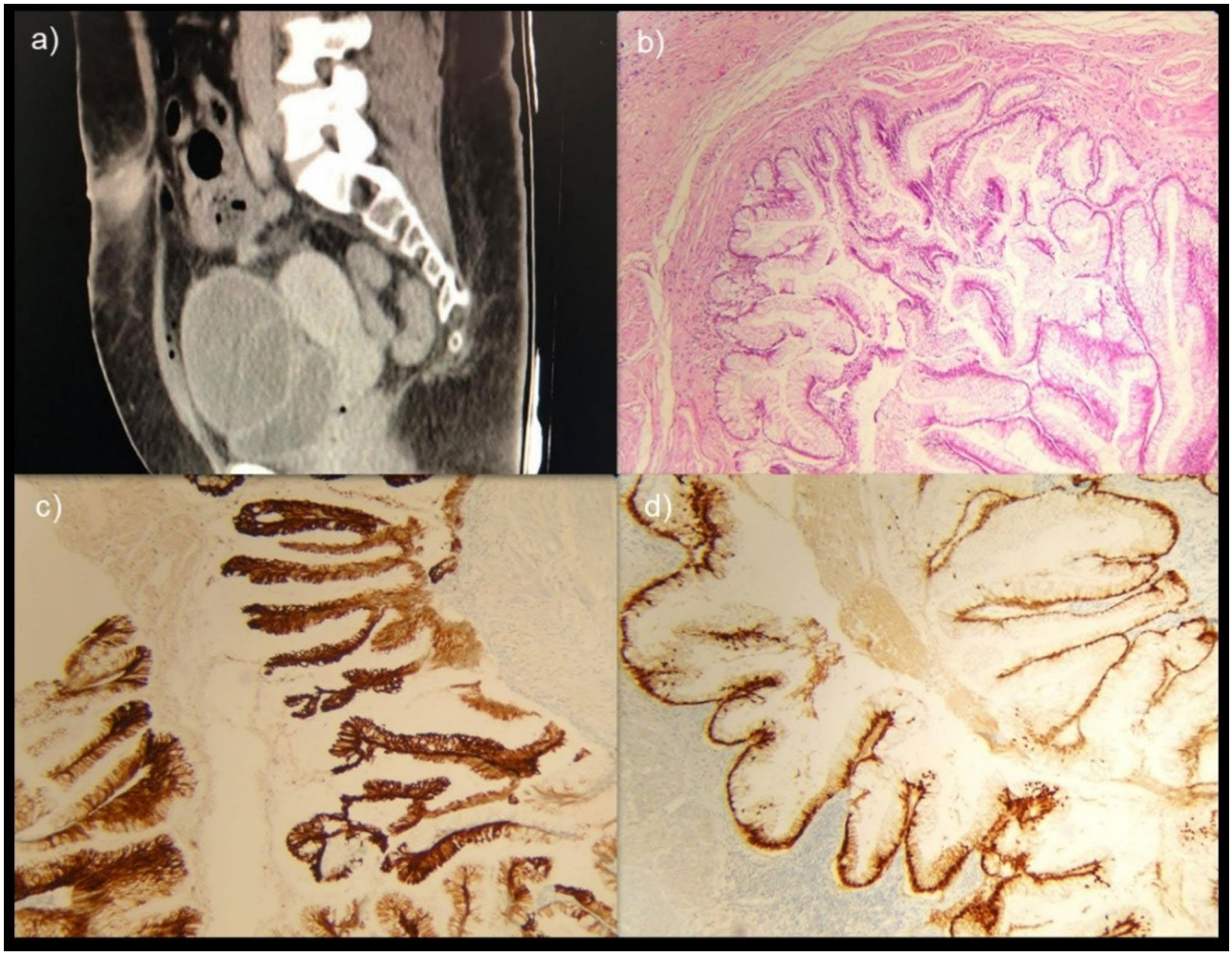

Fig. 1 **a**. An abdominal computerized tomography scan showed a cystic mass of 9 × 8.5 × 8 cm in size in the pelvic region, containing septations and applying pressure to the bladder. **b**. Microscopically, the cyst was lined by a mucinous epithelium that included basally located hyperchromatic nuclei and scattered Goblet cells consistent with an intestinal phenotype. **c**. The tumor that showed papillary formations and mild atypia was positive for cytokeratin 20 immunostain. **d**. The tumor cells were positive for CDX2 immunostain

monitoring (Brennan et al. 2019). Future cases concerning urachal neoplasms should include gross imaging and cystoscopy imaging to contribute valuable data to the medical literature, improve mass characterization, and enhance our overall understanding of these neoplasms. Although data is limited, the prognosis for MCTLMP appears to be excellent following adequate surgical removal. In the case presented, the patient underwent surgical excision, and a follow-up period of 32 months revealed no signs of recurrence or metastasis, further validating the effectiveness of this management strategy.

Given the rarity of MCTLMPs, continued documentation and analysis of such cases are crucial for better understanding their behaviour, prognosis, and potential therapeutic avenues. Additionally, gaining deeper insights into the molecular and genetic characteristics of MCTLMPs may yield valuable information regarding their pathogenesis and potential targets for future therapies.

Urachal MCTLMP's represent an exceptionally rare subset of urachal neoplasms. This case report contributes to the limited body of literature on MCTLMP, highlighting the importance of accurate recognition and diagnosis. The favorable prognosis observed with complete surgical excision further emphasizes the critical role of early intervention in managing MCTLMPs.

In conclusion, this case report sheds light on the rare entity of MCTLMPs originating from the urachus. With only a small number of cases reported in medical literature, this study emphasizes the need for increased awareness and understanding of this unique neoplasm. Histological features, including characteristic cystic architecture with low-grade mucinous epithelial lining and distinct IHC profile, aid in accurate diagnosis and differentiation from other glandular neoplasms.

Surgical excision through partial cystectomy remains the primary treatment option, and long-term follow-up reveals a favorable prognosis following adequate surgical removal. Continued documentation, analysis, and research efforts are vital for gaining deeper insights into the behavior, prognosis, and potential therapeutic targets of MCTLMPs, ultimately improving patient outcomes through early intervention and precise management strategies.

Abbreviations

MCTLMP	Mucinous cystic tumor of low malignant potential
IHC	Immunohistochemical

Acknowledgements
We would like to thank Assoc. Prof. Dr. Esra Paşaoğlu at the University of Health Sciences, Bağcılar Training and Research Hospital for her kind assistance in the diagnosis of this exceptionally rare case.

Authors' contribution
Author YK was responsible for the conception of the case report, acquisition of patient data, and drafting the manuscript. Author MTGM contributed to the writing of the Introduction and Discussion sections, and critically revised the manuscript for important intellectual content. Author FD provided clinical expertise, contributed to the writing of the Case Presentation section, and critically revised the manuscript for important intellectual content. All authors approved the final version of the manuscript and agreed to be accountable for all aspects of the work, ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Funding
The authors did not receive support from any organization for the submitted work.

Data Availability
All data related to the case can be accessed through the following link: https://drive.google.com/drive/folders/1WxcPjk_YiZqywElkTg6AGlZDfdZaKZiD.

Declarations

Consent for publication
The participant has consented to the submission of the case report to the journal.

Received: 2 September 2023 / Accepted: 15 October 2023
Published online: 20 December 2023

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