CASE REPORT

Ruptured Urachal Mucinous Cystic Tumor of Low Malignant Potential: A Case Report and Review of Literature

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Neoplasms of the urachus are uncommon, consisting of only 0.17% of all bladder malignancy. Mucinous cystic tumor of low malignant potential (MCLTMP) is a rare subtype with only 26 cases reported in the literature as of 2023. These tumors may present in a variety of ways such as hematuria, mucusuria, lower abdominal pain and irritative lower urinary tract symptoms. This is a case of 43-year-old female presenting at the emergency room for abdominal pain initially managed as a case of ovarian new growth in complication who underwent exploratory laparotomy, adhesiolysis, bilateral salpingectomy, partial cystectomy enbloc removal of urachal with anterior peritonectomy and excision of umbilicus. Histopathologic examination revealed mucinous cystic tumor of low malignant potential (MCLTMP) of the urachus. It is important to consider the possibility of a ruptured urachal cyst in a female patient who presents with hypogastric abdominal pain. A transabdominal and transvaginal ultrasound may lead to an incorrect diagnosis. In such cases where the patient presented with an acute abdomen, knowledge that a ruptured urachal cyst maybe a differential for such masses would lead to a strategic laparotomy incision aimed at a possible en-bloc removal of the umbilicus while maintaining the connections of the possible urachal mass to the urinary bladder.

Key words: neoplasms of the urachus, urachal tumors, mucinous cystic tumor of low malignancy potential (MCLTMP)

Introduction

Mucinous cystic tumor of low malignant potential (MCLTMP) is a rare subtype of the neoplasms of the urachus. As of 2023, there are only 26 case studies reported in the literature.[1] The case presented as a unique addition to the existing literature as it is the first documented instance of a urachal MCTLMP presenting as acute abdomen with a palpable right lower quadrant mass.

The Case

The patient is a 43-year-old female who presented with a 3-week history of mild hypogastric pain and enlarging abdomen. There were no reported lower urinary tract symptoms or bowel movement changes. She was prescribed pain medications that only temporarily relieved her symptoms. Two weeks after initial treatment, she was rushed to the emergency room due to severe hypogastric abdominal pain. She presented with

a 7cm x 5cm x 5cm palpable right lower quadrant mass with direct and rebound tenderness. Whole abdominal ultrasound revealed a 11.6cm x 10.8cm x 6.8cm ruptured unilocular cyst anterior to the uterus more to the right which was interpreted as a ruptured right adnexal mass probably benign by subjective assessment and IOTA-Adnexa with a 6.25% risk of malignancy (Figure 1A). The right adnexal mass has mixed level echo fluid and multiple echogenic foci within with a point of rupture at the lateral pole measuring 2 cm. It has a capsule measuring 0.4 cm. On Doppler studies, the right adnexal mass shows absent vascularity. (Figure 1B) Transvaginal ultrasound was also done with note of an anteverted uterus with regular contour, cervix with a homogeneous stroma and distinct endocervical canal as well as a uniform and hyperchogenic endometrium. (Figure 1C) She was brought to the operating room with an initial plan of exploratory laparotomy, possible unilateral salpingectomy right. Intraoperatively, a ruptured cystic mass just beneath and adherent to the anterior abdominal wall was noted measuring 13cm x 13cm with egress of yellowish mucinous fluid (Figure 2). The mass had a thick capsule with papillae within. (Figure 4) Upon further exploration, the mass was confluent with the anterior bladder dome (Figure 3). Urology service was then called to assess intraoperatively wherein a suspicion of a large urachal mass was entertained. Since the connections to the umbilicus have already been severed, the surgery proceeded first with a partial cystectomy and en-bloc excision of mass and anterior peritoneum. The umbilicus and the remaining attachments where then excised after. On further evaluation, the patient had normal uterus, normal bilateral adnexae and bowels. The patient had no untoward events post-op and was sent home voiding freely.

Gross pathological examination of the excised mass complex consists of a 25cm x 13.0cm x 6.0cm solitary, tan-brown mass with an attached anterior peritoneum. The external surface of the cystic mass had a grey to tan-brown (13.0cm x 13.0cm x 0.5cm) with dull external surface, located abutting the nearest bladder margin of resection. An oriented rupture point was present on the superior surface of the cystic mass. The internal surface was dark brown and rough with wall thickness measuring

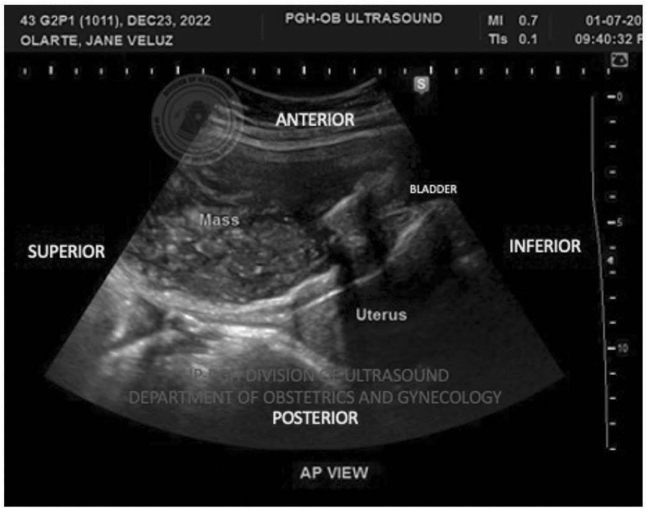

Figure 1A. Holoabdominal ultrasound showing the anatomical relation of the mass, bladder and uterus. The mass is anterior to the uterus more to the right. (UP PGH 2023).

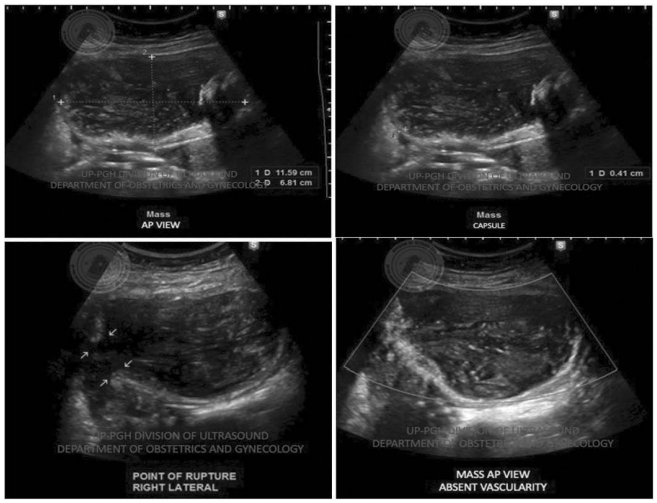

Figure 1B. Holoabdominal ultrasound shows right adnexal mass as a unilocular cyst measuring 11.6cm x 10.8cm x 6.8cm (volume: 445.5 cc) with mixed level echo fluid and multiple echogenic foci within. There are no solid areas and papillary excrescences seen. The capsule measures 0.4 cm. There is a point of rupture measuring 2.0 cm at the right lateral pole of the mass with the contents egressing through the defect. On Power Doppler, the right adnexal mass shows absent vascularity (UP PGH, 2023).

0.3 cm. The cystic mass did not grossly involve the muscularis of attached bladder dome. The oriented anterior peritoneum was grey to dark brown, irregular sheet of tissue (10.0cm x 10.0cm x 1.0cm). The attached bladder dome (6.0cm x 4.0cm x 1.5cm) had a tan-brown, smooth internal surface. The excised umbilicus consisted of a solitary, cream to tan brown, cylindrical, soft to doughy tissue

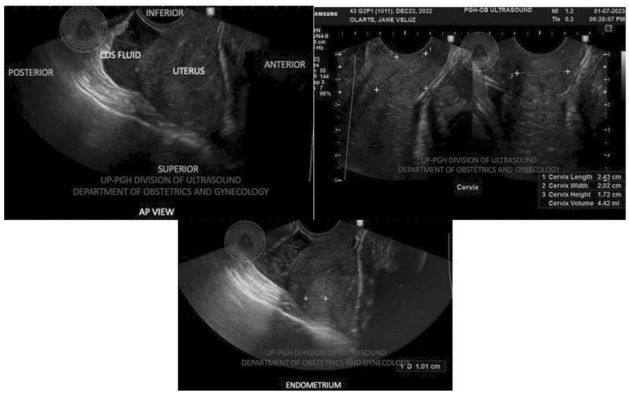

Figure 1C. Transvaginal ultrasound shows The uterus is anteverted with regular contour and homogeneous echopattern measuring 6.9cm x 6.2cm x 3.2cm. The cervix measures 2.4cm x 2.0cm x 1.7cm with homogeneous stroma and distinct endocervical canal. The endometrium is uniform, hyperchogenic, measuring 1.0cm. The endometrial midline is not defined. The endometrial-myometrial junction is regular.

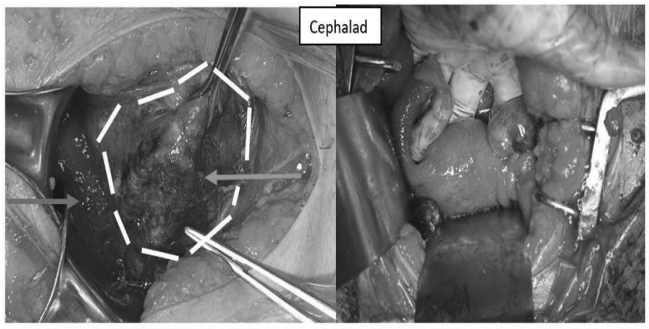

Figure 2. Left 15cm x 10cm cystic mass with egress of jelly-like material (blue arrow) from the point of rupture (Green arrow) Dashed lines show outline of the cystic mass Right. The uterus and bilateral adnexae appear grossly normal.

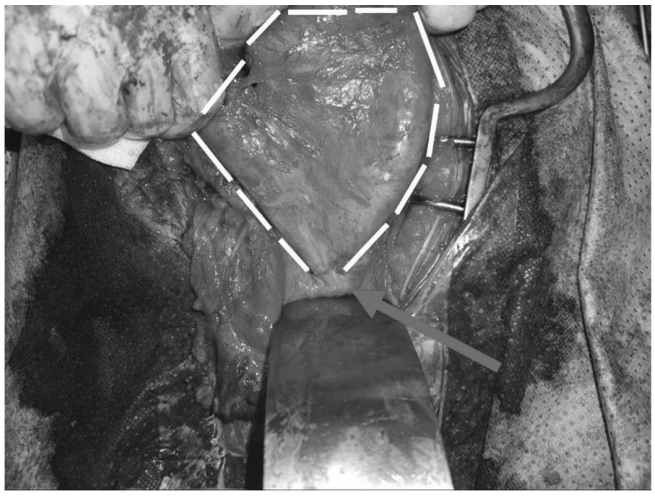

Figure 3. 10cm x 15cm mass (white dashed lines) reflected superiorly with confluence to the dome of the urinary bladder (blue arrow).

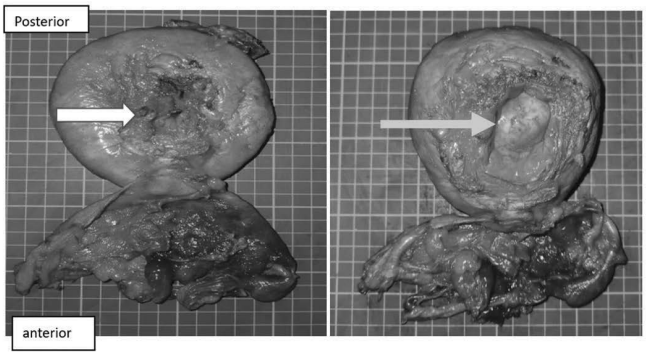

Figure 4. Left The excised mass complex consisted of a 25cm x 13.0cm x 6.0cm solitary, tan-brown bladder mass showing rupture point (White arrow) with an attached anterior peritoneum (Dark arrow). Right- The excised mass complex with attached urinary bladder dome (arrow) UP-PGH, 2023

measuring 4.0cm x 3.0cm x 1.5cm and an attached umbilical skin measuring 2.6cm x 2.0cm x 0.3cm. (Figure 4)

Histologic features of the cystic mass lining show the one to three-layer stratification of the cuboidal to low columnar glandular-type epithelium. Each individual cell has scant eosinophilic cytoplasm, hyperchromatic nuclei with moderate atypia and no definite mitoses. Consistent with the mucinous subtype of the tumor, pools of mucin are prominently seen as the abundant pale eosinophilic stringy material (Figure 5)

The final histopathological diagnosis was urachal mucinous cystic tumor of low malignant potential (MCTLMP). The excised umbilicus and fallopian tubes had benign findings.

A post-operative chest CT scan revealed pulmonary tuberculosis with bronchiectactic and fibrotic changes. No evidence of metastasis was noted. The patient did not receive additional treatment.

At 3 months post-op, she underwent surveillance cystoscopy which showed a smooth urethra and smooth bladder walls with no noted intravesical masses or abnormal mucosal surface. She is on regular follow-up at the outpatient department wherein she is scheduled for periodic imaging with abdominal CT scan with IV contrast.

Discussion

The urachus is a midline tubular structure which is an embryologic remnant of the allantois.

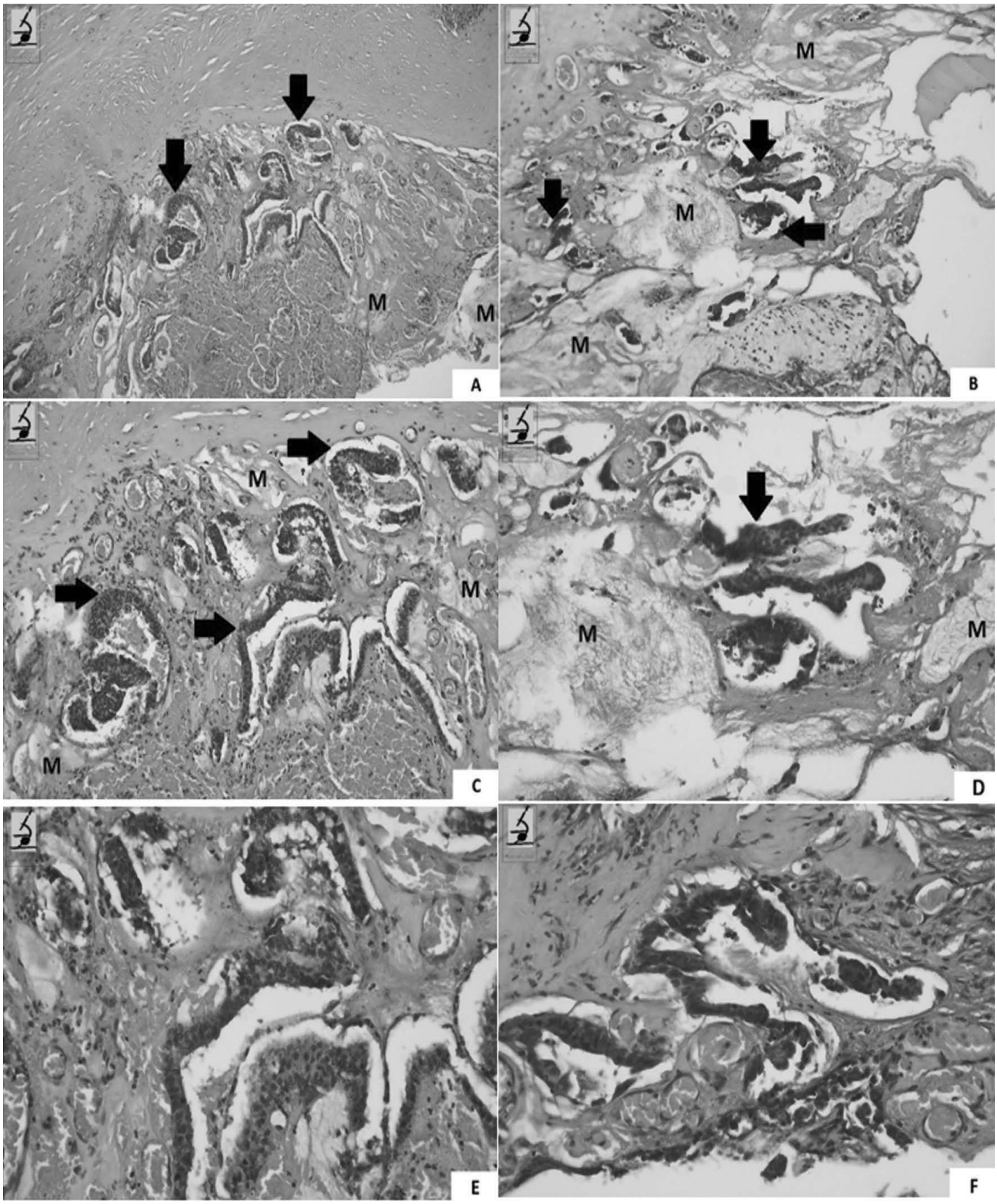

Figure 5. A. and B. (H&E 40X) Internal surface of the cystic mass lined by glandular structures (arrows) in pools of mucin (M). **C**. and **D**. (H&E 100X) Closer magnification of the glands (arrows) and mucin (M). **E**. and **F**. (H&E 400X) High magnification of the glands highlighting its tufted to pseudopapillary architecture, stratification and atypia.

This tubular structure usually involutes before birth or in early infancy forming the median umbilical ligament. However, 32% of adults are reported to have remnant urachus resulting in various clinical presentations such as a patent urachus (most common), umibilical-urachal sinus, vesicourachal diverticulum and urachal cysts.[2] Histologically, the persistent urachus lumen is lined with urothelial epithelium or columnar glandular epithelium. Its walls are formed by smooth muscle in continuation with muscularis propria of the bladder. The exact etiology is not fully understood, but it is believed that the malignant conversion of the columnar or glandular metaplastic epithelium of the urachus may result in the urachal carcinoma.[3]

Neoplasms of the urachus are extremely rare causes of bladder malignancy representing 0.17% of all bladder cancers.[1] There are only a few urachal glandular tumors that are considered completely non-malignant. Villous adenomas and mucinous cystadenomas are examples of completely benign tumors. Villous adenomas present with prominent villopapillary structure, lined by dysplastic mucinous epithelium. Mucinous cystadenomas on the other hand, shows unilocular or multilocular cystic spaces, lined by a single layer of non-dysplastic cuboidal or columnar epithelium.[2]

The mucinous cystic neoplasms are further classified according to the 2016 WHO classification into three entities: Cystadenoma, Mucinous Cystic Tumor of Low Malignant Potential (MCTLMP) and mucinous cyst adenocarcinoma. As of 2023, there are only 48 reported cases of mucinous urachal neoplasms and only 26 cases of mucinous cystic tumor of low malignant potential.[1] The presented case is the first one reported by the institution.

Typically, these tumors manifest with non-specific symptoms like hematuria, mucusuria, urinary frequency, and vague lower abdominal pain. In contrast, some patients exhibited no tumor-associated symptoms, with the tumor being discovered either through palpation or incidentally on imaging. Only two of the reported cases presented with abdominal pain, and merely three cases involved a palpable mass. The case presented is the first documented instance of a urachal MCTLMP presenting as acute abdomen with a palpable right lower quadrant mass. This case stands out in its presentation, thereby enriching the authors' understanding of the variability in clinical manifestations of urachal MCTLMP and highlighting the importance of considering it in differential diagnoses for similar presentations. In a recent review of clinicopathologic characteristics of MCTLMP cases in 2021, it was reported that the incidence is equal between men and women with age ranging from the second to eight decade of life.

The work-up of abdominal masses is one of the highlights of the case. Due to the unusual presentation as well as the failure to recognize the urachal cyst during the initial work-up prior to presenting as an acute abdomen, no cross-sectional imaging like CT scan or MRI was done. Even during transvaginal and abdominal ultrasound done at the ER setting, there was no suspicion of the mass being of urachal in origin. Generally, a midline location is an important feature that helps distinguish an ovarian mass from an urachal mass or bladder mass. Ultrasound may show a mixed echo between the bladder dome and abdominal wall on the umbilical level. However, in the present case, the transvaginal and abdominal ultrasound showed the mass was more to the right and anterior to the uterus. While ultrasonography can demonstrate the tumor as a complex midline mass with a supravesical location, a multiplanar CT or MRI can better differentiate the tumor's anatomic relationship to the nearby structures effectively. Ultrasound is also subject to the operator's technical ability and patient's body habitus.[4]

Abdominal CT scan could reveal a heterogenous, lobulated hypodense mass extending from umbilicus to dome of the urinary bladder wall. Lesions usually show no enhancement after contrast infusion but have internal septa or calcifications of their walls. MRI provides better imaging for the diagnosing the urachus due to its ability to capture detailed images in both coronal and sagittal views, which suit the slanted orientation of the urachus. MRI is particularly useful for showing the spread of disease within the local area or even to other parts of the body. Urachal tumors can appear in various forms—solid, cystic, or both, often containing a jelly-like substance known as mucin. Another common feature of urachal carcinoma is the presence of psammomatous calcifications; fine, round calcifications which are found in most of these cases. Thus, when there is uncertainty on

the location of a pelvic mass, using MRI could be of significant benefit.[5]

First-line and definitive treatment remains to be mass excision, urachectomy or urachectomy combined with partial cystectomy.[6] However, this case was initially assessed as a ruptured ovarian neoplasm, a recognized gynecological emergency, the planning for a possible urachectomy was not considered. An important factor of the case was the failure to identify the urachal cyst during initial evaluation, as discussed earlier. During exploratory laparotomy, the mass was densely adherent to the anterior peritoneum, which hindered further peritoneal dissection and the identification of the bladder. This finding was crucial, as it significantly deviates from the typical presentation of ovarian neoplasms. Even in cases of large ovarian neoplasms, such adherence to the anterior abdominal wall is uncommon. This case illustrates the importance of maintaining a broad differential diagnosis, especially when encountering atypical presentations during surgical procedures. The possible connections of the urachal cyst and the umbilicus was probably dissected during the laparotomy or during further adhesiolysis.

In the literature, there is no reported recurrence or post-operative morbidities noted after mass surgical excision. The prognosis for MTLCP remains good with only 1 mortality described as "death from other causes" and the rest were free of recurrence or metastasis for up to 84 months of follow-up.[1] In contrast to this excellent prognosis, invasive urachal adenocarcinoma has a five-year survival rate 45%.[5] Due to limited data, there is still possibility for further complications, growth, malignant transformation for MCTLMP. Therefore, it is suggested that after surgical excision, periodic imaging and cystoscopy should be done to monitor for recurrence. Cystoscopy and periodic imaging would contribute to the medical literature and increase overall understanding of these neoplasms.[1]

Conclusion

Mucinous cystic tumor of low malignant potential (MCLTMP) is a rare subtype of the neoplasms of the urachus which carries an excellent prognosis. Although rare, the clinician must be familiar with its clinical features and anatomical considerations. This case illustrates the importance of maintaining a broad differential diagnosis, especially when encountering atypical presentations during surgical procedures. Surgical excision, adequate resection and regular follow-up are required to avoid missing the potential malignant component and prevent devastating prognosis.

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