Benign multicystic mesothelioma masquerading as a urachal cyst

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Benign multicystic mesothelioma (BMM) is a benign intra-abdominal lesion that generally occurs in women in their reproductive years. A urachal cyst occurs when the epithelial-lined urachal canal fails to completely obliterate. We report a case of a 38-year-old female presenting with abdominal pain found to have a lesion highly suspicious for a urachal cyst. On pathologic evaluation the lesion was identified as a BMM. This is the first report of BMM presenting as a lesion suspected to be a urachal cyst.

Key Words: benign multicystic mesothelioma, urachal cyst, urachus

Introduction

The urachus is the embryonic remnant of the connection between the urogenital sinus and the umbilicus; ie the allantois. The urachus becomes obliterated in the perinatal period in most people and forms the median umbilical ligament. In some cases when the allantois fails to completely obliterate a span of pathology depending on the degree of failure of obliteration can result including urachal fistula forming a connection from the umbilicus to the bladder, a urachal cyst (the most common type of urachal remnant), a urachal-umbilical sinus or a vesico-urachal diverticulum. Given the risk of development of urachal cancer in these lesions early removal of urachal remnants is generally recommended.[1]

Benign multicystic mesothelioma (BMM) is most commonly an intra-abdominal lesion arising from the peritoneal mesothelium predominantly in woman of childbearing age, though there are reports of this occurring in men and children.[2] BMM arises from the peritoneal mesothelium and can recur after surgical resection with rare reports of malignant transformation.[3] There is also an association of BMM with history of prior abdominal surgery, endometriosis and pelvic inflammatory disease. We report on BMM in a woman of childbearing age with a history of prior abdominal surgery presenting with suspected urachal cyst found to be BMM on final pathologic evaluation.

Urachal remnants and BMM are very different clinical entities that require different management strategies. We present this case of a patient with BMM and review of the current literature and management of this entity.

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Case report

A 38-year-old female with a history of abdominoplasty presented with progressively worse left sided back and abdominal pain. A computerized tomography (CT) scan showed a 3.2 cm x 1.9 cm bilobed fluid filled structure slightly off of the midline to the left near the anterior abdominal consistent with a urachal cyst, see Figure 1. No imaging was done prior to her abdominoplasty for comparison. The patient was given prescriptions for antibiotics and pain medications and on follow up reported improvement in pain, though was still requiring pain medication intermittently. The patient also endorsed history of yellow malodorous discharge from her umbilicus following her abdominoplasty done 8 years prior. She endorsed urinary frequency, urgency, urge incontinence requiring the use of multiple pads per day and sensation of incomplete emptying that had developed over the past couple years. She denied history of urinary tract infections. Her medical history was also significant for diverticulitis, two vaginal deliveries, one cesarean section and history of tubal ligation.

Her examination showed no drainage from the umbilicus. Her abdomen was non-tender and no mass was appreciated. On pelvic examination there was a mass palpated in the left lower quadrant and left adnexal tenderness. Her post void residual was 52 mL. There were no lesions in the bladder on flexible cystoscopy. After discussion of risks of resection including chance that extirpation may not alleviate symptoms the patient elected to undergo surgical management.

At the time of robotic assisted laparoscopic excision of the cystic mass, the lesion was noted to be adherent to the anterior abdominal wall and appeared to be more midline than as noted on the CT scan. Multiple intra-abdominal adhesions were present. The omentum was adherent to the cystic structure. Examination of the bowel and pelvis revealed no gross diverticular disease or endometrial implants. The peritoneum over the anterior abdominal wall was then incised on either side of the median umbilical ligament, the retropubic space was entered and the bladder was dropped. There did not appear to be any communication between the bladder and the cyst. The cyst was excised with wide margins and removed.

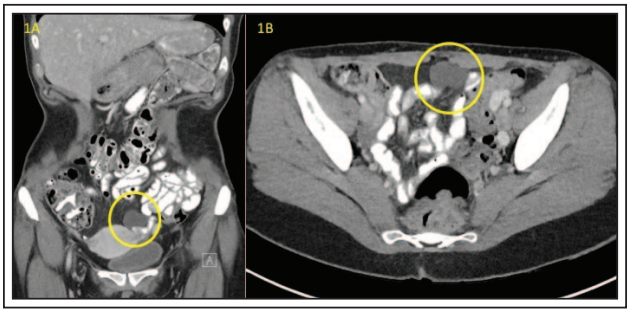

Figure 1. Computerized topographic coronal **(a)** and axial **(b)** images demonstrate a 3.2 cm x 1.9 cm anteriorly located, bilobed fluid filled structure (within the yellow circle) just to the left of the midline concerning for a urachal cyst.

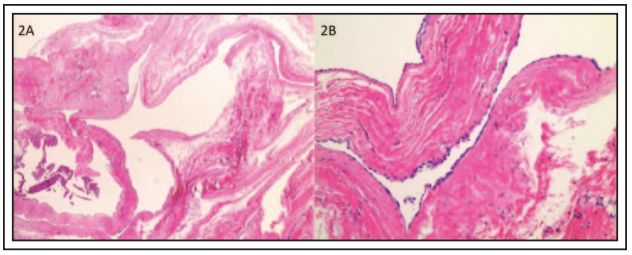

Figure 2. (a) shows a multiloculated cyst with a dense fibrous cystic wall. **(b)** illustrates a single layer of mesothelial cells lining the cyst wall.

The patient did well following surgery and was discharged home on the second postoperative day. Pathology showed a multiloculated cyst with a dense fibrous cystic wall, which was lined with a single layer of mesothelial cells consistent with benign multicystic mesothelioma with negative surgical margins, see Figure 2. On follow up 2 weeks later the patient was pain free and doing well. She was counseled on her diagnosis and will plan to undergo serial imaging with MRI every 6 months given the risk of local recurrence but even more concerning the very rare risk of malignant transformation.

Comment

BMM is a rare tumor with less than 200 reports in the literature. At the time of diagnosis most women are in their reproductive years. The clinical presentation of a patient with BMM varies ranging from shoulder pain, dysfunctional uterine bleeding, dyspareunia, abdominal pain and even bowel obstruction.[4,5] On imaging these lesions can resemble adnexal lesions prompting gynecological evaluation and resection.[5] It has been associated with a history of surgery, pelvic inflammatory disease and endometriosis. BMM is not associated with asbestos exposure. Some theorize BMM is a reactive entity as it is associated with a history of prior abdominal surgery and inflammatory processes.[6]

Less commonly the BMM can present outside of the abdominal cavity including a case involving the tunica vaginalis in an elderly man presenting with scrotal swelling.[7]

Cross-sectional imaging frequently cannot distinguish these lesions from other cystic lesions.[4] The lesions can range in size from several millimeters to 20 centimeters. On cross-sectional imaging these lesions typically appear as a multilocular cystic mass, multiple unilocular thin-wall cysts, or a unilocular cystic mass.[8] Biopsy may help to make this diagnosis prior to surgical intervention. Differential diagnosis generally includes cystic lymphangioma, endometriosis, and adenomatoid tumors. In the current case the appearance mimicked a urachal cyst.

Grossly the cysts are translucent and can be filled with gelatinous or mucinous material. The cysts can be multiloculated or solitary. Under the microscope the cysts are lined with flattened cuboidal or hobnail epithelial cells with exfoliated papillary formation. Ultrastructural examination shows short luminal microvilli, tight desmosomal junctions, extensive rough endoplasmatic reticulum and free ribosomes.[9] Organ invasion has been noted with BMM.

Most agree that cytoreductive approach with wide local excision and peritonectomy is necessary for local control, prevention of recurrence and amelioration of symptoms.[4] There have been some reports of successful treatment with tamoxifen in women. However, only approximately a fifth of cases of BMM are found to be immunoreactive to tamoxifen.[10] Recurrence occurs in approximately 5%0-75% of cases.[4] Malignant transformation has been reported in rare cases.[3] Metastases have not been reported. As cases are rare, there are no evidence based treatment strategies. However, given the risks of recurrence and very small risk of malignant transformation many recommend long term follow up with serial imaging. □

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