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UNUSUAL APPENDICEAL PATHOLOGY PRESENTING AS UROLOGIC DISEASE

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ABSTRACT—We report on 3 cases of unusual appendiceal pathology presenting as urologic disease: 2 cases were benign mucoceles and 1 a malignant mucocele or cystadenocarcinoma of the appendix. Two cases presented as pelvic masses causing urinary frequency and the third with fever and hydronephrosis. The appendix must remain in the differential diagnosis for both acute and chronic disease processes.

The most common pathology associated with the appendix is acute inflammation resulting in peritoneal irritation that if left untreated leads to peritonitis and/or abscess formation and possibly death. Rarer diseases of the appendix include adenocarcinoma, argentaffinoma (carcinoid), benign mucocele, and malignant mucocele (cystadenocarcinoma of the appendix). Malignant mucoceles have a 25 percent chance of rupture into the peritoneal cavity, potentially lining the peritoneum with mucus-producing cells and giving rise to the entity of pseudomyxoma peritonei.[1] Considerable variability in clinical presentation exists because of the mobility of the appendix.[2] Although rare, appendiceal pathology may mimic disorders of the genitourinary tract making the diagnosis difficult even for the experienced surgeon.[3]

Here we report 3 cases of unusual appendiceal pathology presenting as urologic disease. Two cases were benign mucoceles and 1 a malignant mucocele or cystadenocarcinoma of the appendix. Two cases presented as pelvic masses causing urinary frequency and the third with fever and hydronephrosis.

Case Reports

Case 1

A fifty-nine-year-old man presented with worsening urinary frequency, urgency, and pelvic fullness. He denied weight loss, fevers, hematuria, prior abdominal or pelvic surgery, ejaculatory dysfunction, or urinary tract infections. Physical examination revealed a healthy-appearing male with a palpable soft pelvic mass. The mass could be felt on both abdominal and rectal examinations and was considered to be distinct from the bladder and prostate. Abdominal and pelvic computerized tomography (CT) scans confirmed the mass to be separate from the bladder (Fig. 1A). Complete blood count, urinalysis, and urine culture were normal. Initially the patient declined surgical exploration. A serial CT scan showed the mass to be enlarging. The patient's urinary symptoms also worsened. Cystoscopic examination revealed normal mucosa and the trigone had extrinsic posterior posterior wall compression.

Subsequent surgical exploration found a large pelvic mass contiguous with the appendix that was removed en bloc with a portion of cecum as malignancy could not be ruled out at time of surgery (Fig. 1B). However, frozen sections showed inflammatory changes without evidence of neoplasm. The pathologic diagnosis showed acute and chronic appendicitis, organized mucin without evidence of a mucin-producing tumor, and a foreign body giant cell reaction (Fig. 1C). This was consistent with a diagnosis of a benign mucocele of appendix. At six-month follow-up the patient is doing well

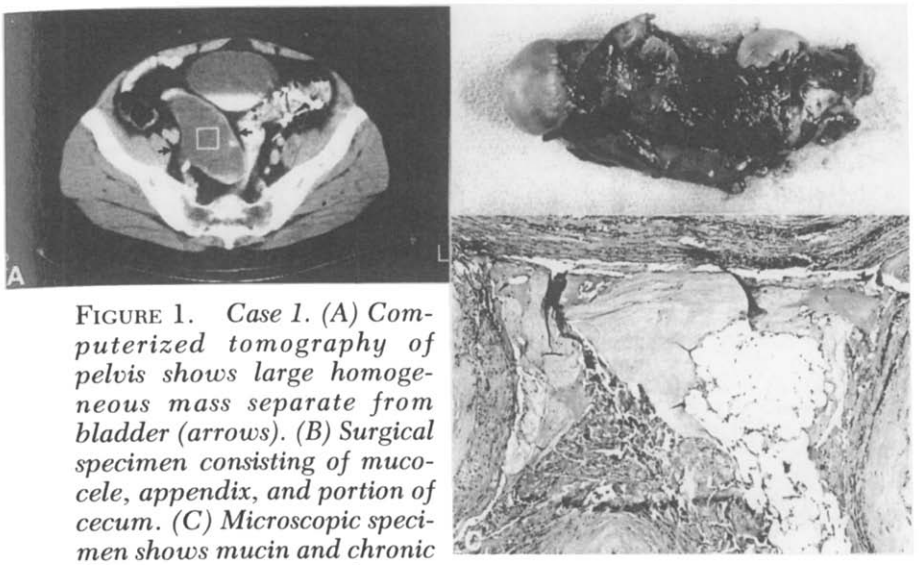
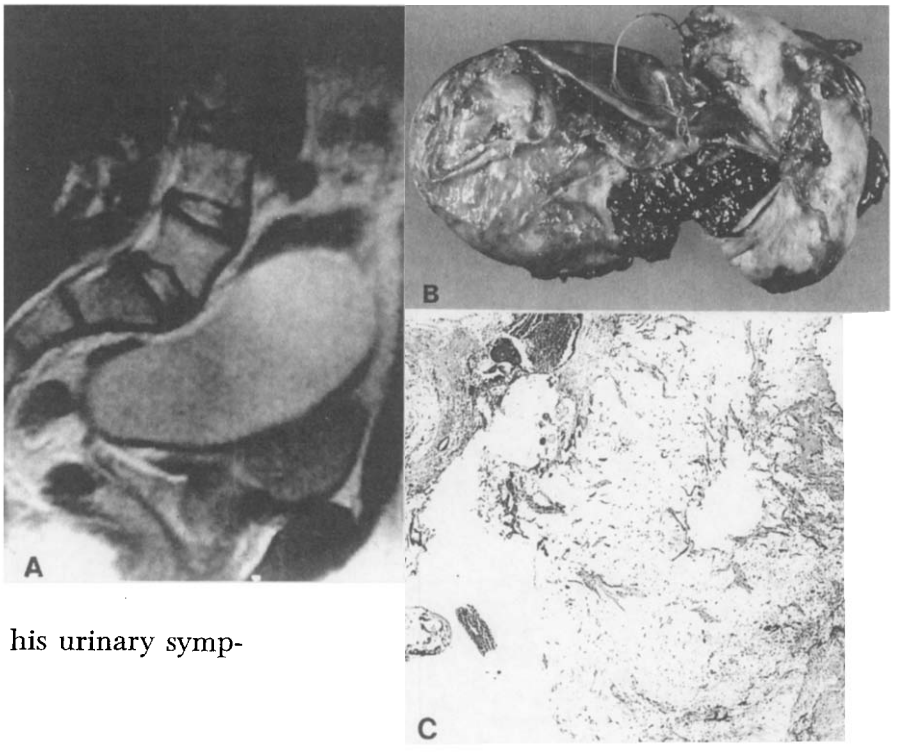


FIGURE 1. *Case 1. (A) Computerized tomography of pelvis shows large homogeneous mass separate from bladder (arrows). (B) Surgical specimen consisting of mucocele, appendix, and portion of cecum. (C) Microscopic specimen shows mucin and chronic inflammation (original magnification × 100).*



2. Case 2. (A) ...etic resonance image of ... shows large mass sepa... from bladder and pros... (B) Surgical specimen ...sting of malignant muco... and appendix. (C) Micro...opic specimen shows mucin ...d atypical mucin-produc... cells consistent with cyst...denocarcinoma of appendix.

with complete resolution of his urinary symptoms.

Case 2

A sixty-four-old man presented for further evaluation of an abnormal prostate examination by his internist. He was asymptomatic without a history of weight loss, urinary tract infection, fevers, or voiding symptoms. He was unmarried and had not fathered any children. Physical examination revealed a healthy-appearing male. Positive finding included a large, soft mass on rectal examination that appeared distinct from the prostate and seminal vesicles. Urinalysis was normal. Initial evaluation included a barium enema followed by pelvic ultrasound, CT scan, and transabdominal cystoscopically directed aspiration, all of which revealed a cystic, midline pelvic mass of uncertain etiology. Also noted was mild bilateral hydronephrosis with normal chemical renal function values.

The patient was lost to follow-up until three years later when he complained of urinary frequency. A repeat CT scan and magnetic resonance image (MRI) (Fig. 2A) showed an increase in the size of the mass along with

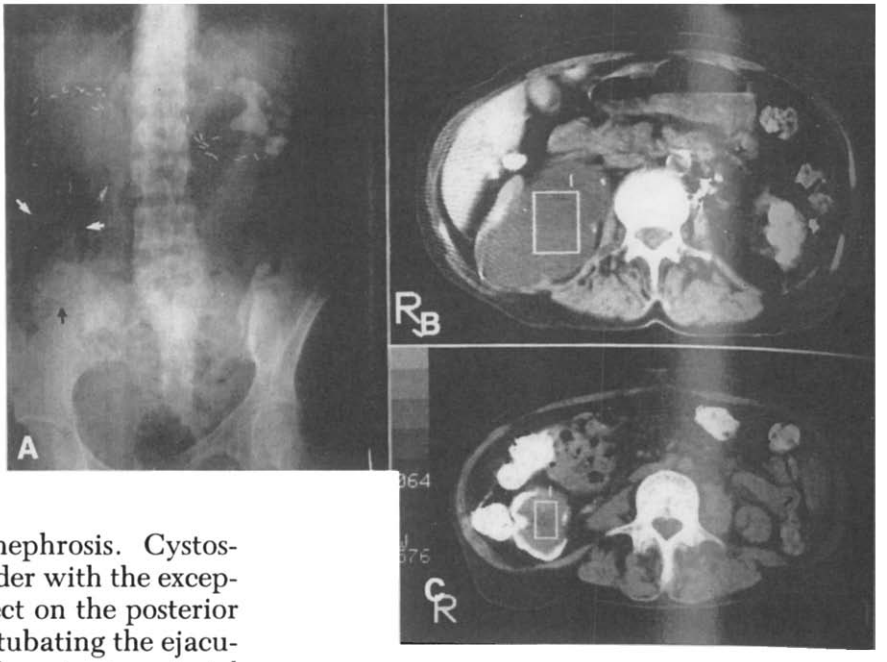


FIGURE 3. *Case 3. (A) KUB film shows left staghorn calculus and calcified mass in right lower quadrant. (B) CT scan of abdomen shows hydronephrotic right kidney with thin rim of renal parenchyma (cursor box in hydronephrotic renal pelvis). (C) CT scan of abdomen shows calcified right lower quadrant mass (cursor box).*

worsening bilateral hydronephrosis. Cystoscopy revealed a normal bladder with the exception of an extrinsic mass effect on the posterior bladder wall. Attempts at intubating the ejaculatory ducts for injection of contrast material were unsuccessful. An exploratory laporatomy was performed at which time a large pelvic mass peeled away from the bladder and anterior rectum (Fig. 2B). Additionally, there was no communication with the prostate. The proximal edge of the mass was contiguous with the tip of the appendix which was demonstrated on opening of the peritoneum. During the dissection the mass was inadvertently opened and a doughy, mucinous, gelatinous tissue spilled and contaminated the peritoneal cavity. An appendectomy was performed along with removal of the mass. Copious irrigation of the peritoneal cavity was performed in hopes of washing away spilled contents. Pathologic analysis of the surgical specimen showed mucin-producing tumor cells consistent with a malignant mucocele or cystadenocarcinoma of the appendix (Fig. 2C). Follow-up at two years showed resolution of voiding symptoms and hydronephrosis as well as no evidence of intra-abdominal disease (pseudomyxoma peritonei) based on a normal CT scan.

Case 3

A sixty-four-year-old woman presented with five days of fever and right flank pain. She was on chronic hemodialysis secondary to bilateral nephrolithiasis and diabetes. Surgical history was remarkable for bilateral renal calculi extraction. Physical examination showed a thin febrile white female. There was right flank and right lower quadrant tenderness. Urinalysis revealed 10 white blood cells per high-powered field with cultures positive for Proteus mirabilis. A film of the kidney-ureter-bladder (KUB) showed a left staghorn calculus and a calcified mass in the right lower quadrant (Fig. 3A). A CT scan was read as severe right hydroureteronephrosis (Fig. 3B, C). A percutaneous nephrostomy tube was placed on the right side with drainage of frank pus. A subsequent nephrostogram showed a grossly dilated right collecting system. The right ureter was not identified. The patient's clinical status did not improve with nephrostomy tube drainage. At surgery a right nephrectomy was performed that revealed a hydronephrotic right kidney. A normal caliber right ureter was found in the usual retroperitoneal location. Interestingly, in close proximity to the ureter was a tubular calcified structure that continued into the cecum where it was found arising from the base of the appendix. The appendix was ligated at the mass removed. Pathology revealed chronic pyelonephritis and hydronephrosis of the right kidney. The appendiceal mass showed mucosa without evidence of mucinous tumor cells. It also showed acute and chronic inflammation with fibromuscular calcifications consistent with the diagnosis of benign mucocele of the appendix. Her postoperative course was uneventful.

Comment

The appendix must be on one's differential diagnosis when evaluating a patient with

TABLE I. *Differential diagnosis of abnormal cystic structures in the male pelvis*[10-13]

Pathology	Location	Characteristics
rial malformation	Lateral	Audible bruit, aneurysm and A/V fistula
dder verticulum	Both	Contiguous with bladder
culatory duct verticulum	Midline	Sperm present on aspiration
dronephrotic lvic kidney	Lateral	Congenital or transplanted
phocele	Both	After pelvic surgery
oceles of e appendix	Both	Contiguous with appendix
llerian duct mnants	Midline	Largest of cystic structures; sperm absent on aspiration
atic cyst	Lateral	Lateral and contiguous with prostate; sperm absent on aspiration
inal vesicle st	Lateral	Associated with renal agenesis; sperm present on aspiration
urethral tula stump	Midline	May cause dribbling incontinence
ffian duct mnants	Lateral	Occur along course vas deferens; sperm absent on aspiration

ute condition of the abdomen. Frequently, do not consider appendiceal pathology hen assessing patients with chronic, long-nding symptoms. Clinical presentation can ary dramatically, due to the inherent mobility of the organ. Appendiceal pathology can mimic umerous urinary tract processes. A recent report, presents 3 cases of proved appendicitis with symptoms suggestive of acute urologic disorders (gross hematuria, acute prostatitis, and acute pyelonephritis).[3] Appendiceal vesical fistula presenting as refractory urinary tract infection is also a well-known entity.[4] Appendicitis mimicking upper urinary tract pathology is also documented.[5] Obstruction of the right ureter with hydronephrosis is known to occur as a result of appendicitis. This can progress to caliceal rupture.[3] Bilateral hydroureteronephrosis also has been reported, the hydronephrosis on the left side thought to be secondary to a functional impairment similar to the paralytic intestinal ileus seen in generalized peritonitis. With appendectomy resolution of the bilateral hydronephrosis was noted.[6]

We herein report 3 unusual cases of appendiceal pathology presenting with urologic manifestations. The symptomatology of 2 patients was caused by large, cystic pelvic masses impinging on the bladder resulting in irritative voiding systems. The differential diagnosis of abnormal cystic, pelvic masses in the male is presented in Table I. Masses can be divided into those that are predominantly midline or lateral in location. Prior surgical history is germane when managing transplanted kidneys and pelvic lymphoceles. It also should be kept in mind, that pelvic structures can be contiguous with, or remnants of the male urologic organs. In both our patients, the urinary tract appeared free from pathology as documented by normal urine sediment, urine cultures, cystoscopic examination and radiologic studies. In hopes of ruling out a müllerian duct remnant in Case 2 intubation of the ejaculatory duct was unsuccessfully attempted. In both cases CT imaging failed to show communication of the mass with the prostate or seminal vesicles. Final diagnosis was made at surgery, which in both patients was self-delayed until increasing urinary symptoms forced intervention.

In Case 1, the final pathology showed a benign mucocele of the appendix. The CT scan (Fig. 1A) preoperatively shows the cyst contents to be homogeneous. Although mucin was found on histologic examination, no evidence of a mucin tumor was noted. The findings of inflammatory cells and foreign body giant cells is consistent with a benign mucocele of the appendix. Symptomatology was related to the size of the mass and its direct impingement on the bladder. Benign mucoceles arise from obliteration of the lumen of the appendix usually from inflammatory scarring or fecaliths. Sterile mucus accumulates behind the obstruction causing progressive cystic dilatation. The cyst may rupture resulting in a local inflammatory reaction.

Case 2 also presented with urinary voiding symptoms secondary to a bladder-compressing, pelvic mass. Pathologic analysis showed mucinous cystadenocarcinoma of the appendix. It is estimated that one fourth of these cases rupture seeding the peritoneal cavity with mucus-secreting cancer cells.[1] This results in multiple mucinous jelly-like implants, i.e., pseudomyxoma peritonei, resulting in morbidity and possible mortality from adhesions and bowel obstruction. Interestingly, these cells rarely invade the underlying peritoneal wall hence visceral metastases are uncommon.[1] In females, pseudomyxoma peritonei is usually from the intra-abdominal spread of a ruptured cystadenocarcinoma of the ovary.[7] In males, the most common etiology is from a mucinous cystadenocarcinoma of the appendix as seen in Case 2.[8] Typically, pseudomyxoma peritonei presents as vague abdominal pain or if the disease is more extensive, weight loss, constipation, and fatigue.[9] In our patient, the tumor was localized

to the appendix without any intraperitoneal deposits. Of concern was the inadvertent leakage of mucinous material into the peritoneal cavity during surgical removal. Close follow-up has shown no signs or symptoms of pseudomyxoma peritonei and at two years of follow-up a CT scan of the abdomen and pelvis was normal.

In Case 3, the final pathology showed a benign mucocele of the appendix. This patient had chronic urinary stone disease and a clinical presentation consistent with right pyonephrosis. In retrospect, the KUB (Fig. 3A) and CT scan (Fig. 3B, C) of the abdomen reveal a calcified mass on the right side consistent with a mucocele. The right kidney has only a thin rim of parenchyma (Fig. 3B arrow) and was without sufficient function and/or was obstructed so that the right ureter was not readily apparent on the CT scan. The hydronephrotic kidney and mucocele also have slightly different Hounsfield units on the CT scan (Fig. 3B). The etiology of the hydronephrosis was most likely a combination of mucocele obstruction and previous nephrolithiasis. The close proximity of the right ureter and appendix can lead to both pre- and intra-operative confusion. An appendiceal fecalith can also mimic ureteral calculi. Here, chronic appendicitis was mistaken for a dilated calcified ureter.

In conclusion we report 3 cases of unusual appendiceal pathology that presented as urologic disease. All 3 cases were successfully managed with appendectomy and mucocele excision. The appendix must remain in the differential diagnosis for both acute and chronic disease processes.

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