Current management of the urachal anomalies (UA). Lessons learned from the clinical practice

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Abstract

Purpose It has been suggested that symptomatic UA requires surgical excision. However, the management of asymptomatic urachus is still controversial. We aimed to evaluate the clinical presentation, the efficacy of current modalities used, and postoperative pathology in patients with UA.

Materials and methods We have performed a retrospective review of all patients diagnosed with UA and treated surgically or conservatively over 18 years. Demographic data, clinical presentation, imaging modalities, pathology, treatment, and postoperative complications were analyzed.

Results Twenty-five symptomatic patients (18 males and seven females) with a median age of 13 years (1 month to 37 years) were identified. 15 (60%) were diagnosed with a urachal cyst, 4 (16%) with sinus, 3 (12%) with urachal diverticulum, and the remaining 3 (12%) with patent urachus.

Of those, 20 (80%) underwent surgical repair, and the remaining five (20%) patients were managed conservatively. 4 (20%) underwent laparotomy, 7 (35%) laparoscopic incision, and the remaining 9 (45%) laparoscopic robotic-assisted surgery. Nine patients required bladder cuff excision. The median operative time was 75 min (42–140 min).

One patient developed Clavien–Dindo grade IIIA complication resulting in infected hematoma, which resolved after drainage. Another patient with a complication of grade IIIB needed reoperation as a result of recurrent events of an abscess. 13 (65%) demonstrated epithelium lining of the urachus on postoperative pathology.

Conclusions Our data show that most of the patients with UA presented with epithelial lining, which might lead to the later malignant transformation. It might cause a shift from the conservative management of asymptomatic patients to surgical intervention. Robotic-assisted surgery appears beneficial in these patients, especially when the bladder cuff excision is required.

Keywords Urachus anomalies · Robotic-assisted surgery · Urothelium · Ultrasound

Introduction

The urachus is an embryologic remnant, a midline tubular structure length of 3–10 cm that extends from the anterior dome of the bladder toward the umbilicus [1]. Urachus arises from allantois separation from the ventral cloaca. The bladder descends into the pelvis, and as a result, the urachus gets loose shortly after birth. The tubular structure is obliterated and changed into a fibrous cord called the Median umbilical ligament. The failure of this process establishes a remnant of the urachus [2, 3]. It’s a rare congenital anomaly, with an incidence of 1.6% under 15 years old and in adults is 0.063% [4]. Urachus anomalies (UA) are more common among males and are subclassified into four congenital pathologies: 1. Urachal sinus (no connection with the bladder but the pouch open to umbilicus), 2. Patent urachus (communication

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between the umbilicus and bladder), 3. A urachal cyst (no connection to either bladder or umbilicus, the central part filled with fluid), 4. Urachal diverticulum (pouch open to the dome of the bladder) [5, 6]. It has been reported that urachal cyst is the most common type with 45% of cases, followed by urachal sinus with 37%, patent urachus in 16%, respectively [6]. There’s no consensus about the urachal anomalies management approach [7, 8]. Recent studies support the hypothesis that symptomatic and asymptomatic children will benefit from performing surgical excision with a bladder cuff because of the risk of malignant transformation [9]. However, some have emphasized that unnecessary surgeries should not be performed since the malignant transformation is very rare, therefore advocating a more conservative approach. The surgical procedure could be performed utilizing several approaches, including open, laparoscopic, or robotic assist. Urachal adenocarcinoma in childhood is not reported yet, but transformation can occur in adult life and present with high mortality and more surgical complications [10]. We have aimed to evaluate the clinical presentation, the efficacy of modalities used, and the comparison between management approaches and postoperative pathology to establish recommendations for daily clinical practice.

Materials and methods

A retrospective review of electronic records from the departments of Pediatric urology and Surgery was performed on all patients diagnosed with UA and treated surgically or conservatively between the years 2002 to 2021. Patient information consisting of age, gender, clinical manifestation, imaging modalities, type of anomaly, treatment approaches, postoperative complications, and postoperative pathology were evaluated.

Results

A total of twenty-five patients were identified with UA at our institution during the study period. The demographic data on the studied patients are presented in Table 1. In brief, the median age was 13 years old (1 month to 37 years old), 72% were males, and 28% were females. The urachal cyst was diagnosed in 15 (60%), urachal sinus in 4 (16%), urachal diverticulum in 3 (12%), and patent urachus in 3 (12%), respectively. All patients were symptomatic at presentation. Umbilical discharge was in 15 (64%), abdominal pain in 6 (24%), recurrent urinary tract infection in 2 (8%), hematuria in 2 (8%), and enuresis in (8%), respectively. The diagnostic modalities used for initial evaluation were abdominal ultrasound (US) in 22 patients, CT scan in 8, MRI scan in 2 respectively, and four underwent Voiding Cystourethrogram (VCUG). The US was utilized for the initial and final diagnosis in 88% of patients. CT scan was performed on eight patients; in five US demonstrated the same results. In two patients, MRI was utilized; however, the MRI finding were identical to the US. In one patient US was reported as normal, but urachal diverticulum was demonstrated on VCUG.

Five (20%) out of 25 patients were discharged for conservative follow-up since all clinical symptoms were resolved and the parents decided to proceed with the conservative treatment. They disappeared from the follow-up. Twenty patients (80%) underwent surgical intervention. Of those, four (20%) underwent laparotomy with an average time of 110.2 min (range 93–145 min), seven (35%) laparoscopic excision of the UA with an average time of 66.4 min (range 50–107 min), and nine (45%) laparoscopic robotic-assisted excision with an average time of 79.5 min (range 52–140). Of those patients, who underwent a robotic-assisted excision, 5 (55%) have had bladder cuff excision. Postoperative complications were occurred in 2 (11.1%) patients, one patient after open surgery had class IIIB, and another patient after Robotic-assisted surgery had class IIIA, according to the Clavien–Dindo classification. On pathological examination of the excised urachus, urothelium (Fig. 1.) was present in 13 (65%) out of 20 samples. Of those in three, necrotizing granulomas were revealed. Additionally, in one patient urachal duct was lined by mucus-producing epithelium with goblet cells and without gastrointestinal epithelium. No epithelium cover was found in 7 (35%) out of the twenty patients; six have had obliterated urachus, and the other one demonstrated cystic fibrous walls without epithelium.

Discussion

This study ironed out the previously published data demonstrated predominant incidence of UA in the younger patients with higher frequency among males. UA is often found incidentally on imaging, although in our case series, 100% of patients were symptomatic at the time of diagnosis and umbilical discharge was the most common presentation (64%) [11]. Additional complications of urachal anomalies such as progressive growth, stones, intracystic bleeding, intraperitoneal rupture, bowel fistula, bowel obstruction, and malignancy are rare, as also we have observed in presented here study [11]. The diagnosis of UA in this study was confirmed by varying radiographic methods; however, the US was an initial and final method in the majority of the cases. Other modalities were used, such as CT or MRI, however, they just confirmed US findings. VCUG was utilized in a few of our patients; in one patient, the US had unremarkable, and the VCUG provided an accurate diagnosis of a urachal diverticulum. This is inconsistent with the series of M.O.
In our opinion, US is the best diagnostic tool as an initial diagnostic method; in patients in which the US is inconclusive, the subsequent imaging with CT or MRI scan is required. VCUG could be considered when the clinical presentation suspect UA and other imaging methods have had no significant value. There are still conflicting opinions regarding therapeutic options for optimal UA management. Several authors suggest that asymptomatic incidentally diagnosed urachal anomalies are normal variants that don’t need any intervention with a high chance for spontaneous resolution, regardless of UA type [2, 7, 8]. On the other hand, some argue that prophylactic surgery should be utilized in all patients in order to prevent progression to the symptomatic UA and further complications such as malignant transformation [5].

In our case series, 80% of patients underwent surgery regardless of the UA type or whether the infected UA is present, as other authors have suggested [5]. We

| Patient | Sex | Symptom | Diagnosis | Treatment + bladder cuff excision | Complication Pathology |
|---------|-----|---------|-----------|-----------------------------------|------------------------|
| 1       | M   | Discharged + enuresis | US | Conventional laparoscopy | NA | Urachal duct, partially obliterate lined by mucin-producing epithelium with goblet cells |
| 2       | M   | Local infection + discharged | US | Robotic-assisted | NA | Small duct with urothelium |
| 3       | M   | Local infection | US + VCUG | Robotic-assisted | NA | Small duct with urothelium |
| 4       | M   | Local infection | US | Conventional laparoscopy | NA | Obiterated urachus |
| 5       | F   | Local infection + discharged | US | Laparotomy + bladder cuff excision | NA | Small duct with urothelium and necrotizing granuloma |
| 6       | M   | Local infection | US | Robotic-assisted | NA | Small duct with urothelium |
| 7       | M   | UTI + hematuria + enuresis | US + VCUG | Conventional laparoscopy | NA | Small duct, partly obliterated with urothelium epithelium |
| 8       | F   | Local infection | US + MRI | Laparotomy + bladder cuff excision | IIA- Drainage | Small duct with urothelium |
| 9       | M   | Discharged | US | Robotic-assisted + bladder cuff excision | NA | Cystic fibrous wall without epithelium |
| 10      | F   | Local infection + abdominal pain | US + MRI | Robotic-assisted + bladder cuff excision | NA | Small duct with urothelium |
| 11      | M   | Local infection + abdominal pain | US | Robotic-assisted + bladder cuff excision | NA | Obliterated urachus |
| 12      | F   | Local infection + abdominal pain | US | Laparotomy + bladder cuff excision | NA | Small duct with urothelium and necrotizing granuloma |
| 13      | F   | Hematuria | CT | Laparotomy + bladder cuff excision | NA | Urachus cyst covered by urothelium |
| 14      | M   | Discharged + abdominal pain | CT | Robotic-assisted + bladder cuff excision | NA | Small duct with urothelium and necrotizing granuloma |
| 15      | F   | Discharged | CT | Conventional laparoscopy | NA | Obliterated urachus |
| 16      | M   | Discharged + local infection | US + CT | Conservative | NA | NA |
| 17      | M   | Discharged + local infection | US + CT + VCUG | Conventional laparoscopy | NA | Obliterated urachus |
| 18      | M   | Discharged + local infection + abdominal pain | US + VCUG | Conservative | NA | NA |
| 19      | M   | Discharged + Local infection + abdominal pain | US + CT | Conservative | NA | NA |
| 20      | M   | Discharged | US + CT | Conventional laparoscopy | NA | Obliterated urachus |
| 21      | F   | Discharged | US + CT | Conventional laparoscopy | NA | Obliterated urachus |
| 22      | M   | Discharged + local infection | US | Conservative | NA | NA |
| 23      | M   | Discharged + local infection | US | Conservative | NA | NA |
| 24      | M   | Discharged + UTI | US | Robotic-assisted | NA | Small urachus duct with urothelium |
| 25      | M   | Abdominal pain | US | Robotic-assisted + bladder cuff excision | NA | Small urachus duct with urothelium |

McCollum et al. in which VCUG has been misleading and added no relevant data to ultrasound studies [11].
have used different surgical approaches along evolution of the surgical techniques and technology. Although laparoscopy is the method of choice in the majority of the centers, we prefer to use a robotic-assisted surgery [12]. The anatomical details are better visualized utilizing the robotic-assisted method; furthermore, robotic intracorporeal suturing is easy during bladder cuff excision and bladder closure when required (Supplementary file). Our complication rate was comparable and even superior to the previously reported in the literature. Only one patient developed Clavien Dindo grade IIIA complication resulting in infected hematoma, which was transcutaneous drainage. Another patient had a complication grade IIIB resulting in a recurrent event of an abscess, which needed reoperation. Naiditch et al. reported a 14.7% complication rate, with all needed re-operation. In the reported McCol- lum et al. series of 26 patients, one patient had a wound infection, and one had a bladder leak. Surgical excision is aimed to prevent infectious and malignancy. Urachal carcinoma is rare; it accounts for less than 1% of bladder cancer, however, it is very aggressive, with a low survival rate of less than 50% [13]. In our case series, we wanted to examine the relationship between the patient’s clinical presentation and the histopathology features in adult and pediatric urachal anomalies in order to better understand who can be allocated to conservative treatment and who could benefit from a surgical procedure. Malignancy arises from epithelial cells; therefore, we could speculate that epithelium findings at the site of the excised UA would be a high-risk potential for future malignant transformation. Overall, 13 (65%) of our 20 cases contained urothelium. Three (23%) out of the 13 were found to have urothelium with necrotizing granuloma. One patient has had a distributing pathology, such as mucin-producing epithelium with goblet cells and without gastrointestinal epithelium. This finding placed him to the group of patients with a higher potential risk for further malignant transformation [13].

In contrast, remnants without epithelium had a low prediction for urachal cancer. Thirty-five percent of studied here patients appeared with no epithelium, and of those 6 out of 7 patients had obliterated urachus. We found no association between the symptoms and the ability to predict the pathology. Copp et al. suggested that asymptomatic patients might present epithelium elements as symptomatic patients [14]. Hence, due to the difficulty in predicting the pathology based on the clinical presentation, we recommend a surgical procedure for all patients with UA in order to prevent future complications regardless of clinical presentation.

The limitations of our study should be mentioned. It is a retrospective study and suffers all flaws of such kind. There is a lack of complete information data from the medical records, a relatively small study number. Additionally, there is a lack of uniform follow-up imaging and detailed information. However, the follow-up was meticulous in studying here patients, and the same pathologist (SZ) with fellowship training in UG pathology evaluated surgical specimens over the all study period, so no bias was presented.

Conclusions

Our data show that most patients with UA have epithelium presence. We are not able to predict the pathology based on clinical manifestation. Therefore, we recommend performing surgical removal of UA regardless of symptoms. Our method of choice for these patients is robotic-assisted surgery, which provides a better outcome, especially when bladder cuff excision is needed.

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Declarations

Conflict of interests All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

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