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Urachal Anomalies in Children: A Single Center Experience

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The objective of this study is to define optimal diagnosis and treatment strategies for patients with urachal anomalies in the pediatric age group. The medical records of 21 children who had undergone surgery for urachal anomalies at Severance Hospital, Yonsei University College of Medicine from January 1990 to April 2005 were reviewed. The subjects included 14 males and 7 females (M:F 2:1). The four types of urachal anomalies confirmed were a urachal cyst in 10 patients (47.6%), a patent urachus in 6 (28.6%), a urachal sinus in 4 (19.0%) and a urachal diverticulum in 1 (4.8%) patient. The most common presenting complaint was umbilical discharge (n = 10, 40.0%), followed by abdominal mass (n = 9, 36.0%). Urachal anomalies were diagnosed by ultrasonography in 18 patients, and 7 of them were additionally examined by computed tomography. The remaining patients were diagnosed solely by surgical exploration. Excision was performed in all patients and was supplemented by partial cystectomy in three. Umbilical discharge was the most common clinical manifestation in our patients, suggesting that ultrasonography should be performed in patients with umbilical discharge to differentiate urachal anomalies. We found the most common anomaly to be the urachal cyst, and all patients were successfully treated by surgical excision.

Key Words: Urachal anomaly, children, ultrasonography, surgical excision

INTRODUCTION

The urachus, or median umbilical ligament, is a normal embryonic remnant of the primitive bladder dome and extends upward from the anterior dome of the bladder toward the umbilicus.[1,2] It generally exists as a fibrous vestigial remnant of at least two embryonic structures: the cloaca, which is the cephalic extension of the urogenital sinus (a precursor of the fetal bladder), and the allantois, which is a derivative of the yolk sac.[3] The tubular urachus is normally changed into a fibrous band after birth. If this regression is incomplete, disorders of the urachus can appear. Urachal remnant diseases are rare and manifest with nonspecific abdominal or urinary signs and symptoms. Although rare, these anomalies can present with acute life-threatening infection[4-6] and have commonly been associated with malignant degeneration.[7-9] For these reasons, it is important for the pediatrician to formulate an accurate diagnosis and proper treatment. However, definitive presurgical diagnosis is not easily made because of the low prevalence of urachal anomalies.

The aim of this study is to define optimal diagnostic and treatment strategies for patients with urachal anomalies in the pediatric age group.

MATERIALS AND METHODS

The medical records of a total of 21 children with urachal anomalies who were treated at Severance Hospital, Yonsei University College of Medicine from January 1990 to April 2005 were retrospectively reviewed. The patient records were reviewed with regard to age, sex, clinical presentation, diagnostic studies and their accuracy, type of urachal anomaly, associated anomalies, microbiology, surgical management and complications. All of the patients had undergone surgical excision. The urachal anomalies were

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classified into four types according to their anatomical structure as identified during surgery.[10] They included urachal cyst, patent urachus, urachal sinus, and urachal diverticulum.

The diagnosis of a urachal anomaly was established only by history and physical examination in 3 of the 21 children: 2 with a patent urachus and 1 with a urachal sinus. Radiographic studies were performed to diagnose urachal anomalies in the remaining 18 children. Ultrasonography, computed tomography, voiding cystourethrography, and fistulogram were used.

RESULTS

Twenty-one patients with urachal anomalies were identified; there were 14 boys and 7 girls, giving a 2:1 ratio of males to females. The mean age was 3.9 years (range: 1 day to 12 years). Urachal anomalies were most common in infancy, from one day to two years of age (42.9%). Preschool children from two to six years of age accounted for 33.3%, while children from 6 to 12 accounted for 23.8% (Table 1). The patients' symptoms are summarized in Table 2. Umbilical discharge was the most common symptom (40.0%) followed by abdominal mass (36.0%). As shown in Table 3, there were four types of urachal anomalies including urachal cyst, patent urachus, urachal sinus and urachal diverticulum. A urachal cyst, which was most common, was diagnosed in 10 patients (47.6%). Several associated anomalies are umbilical hernia, inguinal hernia, omphalomesenteric duct sinus, bowel fistula, TOF, and ASD2.

All eighteen patients with a final diagnosis underwent ultrasonography and additional radiographic studies such as computed tomography, voiding cystourethrography and fistulography. Patients with a final diagnosis of a urachal cyst had undergone 10 ultrasonographs (7 diagnostic) and 3 computed tomograms (2 diagnostic). Patients with a final diagnosis of patent urachus had four ultrasonographs (1 diagnostic), 3 computed tomograms (1 diagnostic), a single diagnostic voiding cystourethrography and a single diagnostic fistulograph performed. Patients diagnosed with urachal sinus had undergone 3 ultrasonographs (2 diagnostic) and a single diagnostic computed tomogram, while the patient with urachal diverticulum had undergone a single diagnostic ultrasonograph (Table 4). The remaining three patients were diagnosed only through surgical exploration because their symptoms strongly indicated the presence of urachal anomalies.

All patients were successfully treated by surgical excision. Among them, three patients also underwent partial cystectomy. There were no postoperative complications and recurrences after treatment. The urachal anomalies of nine patients were suspected to be infected and samples were cultured. Staphylococcal species were isolated in two patients, Peptostreptococcus in one, Streptococcus in one, and Escherichia coli in one patient. Cultures of other patients had no growth.

DISCUSSION

According to Begg, the persistence of the urachus was described and treated for the first time in 1550 by Bartholomaeus Cabrolius.[11] The urachus is a lower midline abdominal structure that extends from the dome of the bladder toward the umbilicus, as the median umbilical ligament

Table 1. Age and Sex Characteristics of Urachal Anomalies

Age (yrs)	Sex		Total (%)
	Male	Female	
< 2	6	3	9 (42.9)
2 - 6	5	2	7 (33.3)
6 - 12	3	2	5 (23.8)
Total	14	7	21 (100)

within the space between the peritoneum and transversalis fascia; it is also known as the space of Retzius.[2,10] Urachal remnants are an end result of incomplete regression of the intra-embryonic connection between the allantois and the cloaca. The allantois appears on about day 16 as a tiny, fingerlike outpouching from the caudal wall of the yolk sac, which is contiguous with the ventral cloaca at one end and the umbilicus at the other. The ventral portion of the cloaca develops into the bladder after cloacal division by the urogenital septum. Thus, the bladder initially extends all the way to the umbilicus.[12] The exact timing of urachal closure is uncertain, but it is generally thought that the bladder descends into the pelvis and that its apical portion progressively narrows to a small, epithelialized, fibromuscular strand by the fourth or fifth month of gestation. The urachus is then completely obliterated to a thin fibrous cord by birth.[3] The urachus varies from 3 to 10 cm in length and from 8 to 10 mm in diameter. It is a three-layered tubular structure, the innermost layer being lined with transitional epithelium, the middle layer composed of connective tissue, and the outermost muscular layer in continuity with the detrusor muscle.[13]

The incidence of urachal anomalies is low. Pediatric autopsy studies have shown an incidence of 1 in 7,610 cases of patent urachus and 1 in 5,000 cases of urachal cysts.[14] Drawson et al. reported 2 cases in 300,000 hospital admissions and Nix et al. reported 3 cases in 200,000 hospital admissions in Boston and 3 cases in 1,168,760 hospital admissions in New Orleans.[15,16] As shown by previous differences between autopsy cases and hospital admission cases, urachal anomalies are not easily encountered in clinical cases because they are

Table 3. Types of Urachal Anomalies

Type	Cases (%)
Urachal cyst	10 (47.6)
Patent urachus	6 (28.6)
Urachal sinus	4 (19.0)
Urachal diverticulum	1 (4.8)
Total	21 (100)

Table 2. Clinical Manifestation

Symptom & sign	Urachal cyst	Patent urachus	Urachal sinus	Urachal diverticulum	Total
Umbilical discharge	0	6	4	0	10
Abdominal mass	7	1	0	1	9
Abdominal pain	2	1	0	0	3
Fever	1	0	0	0	1
Dysuria	0	0	1	0	1
Frequency & nocturia	1	0	0	0	1

Table 4. Diagnostic Studies Performed

Test	Urachal cyst		Patent urachus		Urachal sinus		Urachal diverticulum	
	Performed	Diagnostic	Performed	Diagnostic	Performed	Diagnostic	Performed	Diagnostic
US	10	7 (70.0%)	4	1 (25.0%)	3	2 (66.7%)	1	1 (100%)
CT	3	2 (66.7%)	3	1 (33.3%)	1	1 (100%)	0	0
VCUG	1	0	1	1 (100%)	1	0	0	0
Fistulogram	1	0	1	1 (100%)	0	0	0	0

US, ultrasonography; CT, computed tomography; VCUG, voiding cystourethrography.
Diagnostic accuracy: US: 61.1%, CT 57.1%.

frequently asymptomatic. The incidence rate in males is twice as high as in females.[17] This study showed the same result with 14 boys and 7 girls having urchal anomalies. Urachal anomalies were most common in infancy ranging from one day to two years of age. The majority of urachal anomalies can be classified into one of the following groups: patent urachus (most common, 48%), in which the entire tubular structure is intact; urachal sinus (18%), in which the umbilical end of the structure fails to close; urachal diverticulum (3%), in which the bladder end of the structure fails to close; urachal cyst (31%), in which both ends close but the central lumen remains open; and alternating sinus, which is a cyst-like structure that can drain to either the bladder or the umbilicus.[10]

The reported incidence rates of these five anomalies have varied among different studies. In our series, four types were identified; urachal cyst, patent urachus, urachal sinus and urachal diverticulum. A urachal cyst in 10 patients (47.6%) was the most common diagnosis in this study. A patent urachus in 6 patients (28.6%), a urachal sinus in 4 (19.0%) and a urachal diverticulum in 1 patient (4.8%) followed. We suspect that the differences in rates between our study group and the common classification group stems from the small number of cases we investigated.

Urachal anomalies present various clinical manifestations. A urachal cyst may sometimes occur as an abdominal mass and develop into an abscess in the abdominal wall if infected. A patent urachus often presents early in infancy as clear drainage from the umbilicus, but the drainage may contain blood or pus if infected. As for an urachal sinus, symptoms may include generalized pain, fever, periumbilical pain, periumbilical redness or drainage. A urachal diverticulum is often asymptomatic and is usually found incidentally during investigations for other problems. A urachal diverticulum can often be associated with recurrent urinary tract infection. An alternating sinus can empty either into the bladder or the umbilicus and this characteristic can cause it to present various manifestations.[18]

A thorough history and physical examination is necessary for the diagnosis of urachal anomalies. The definitive diagnosis requires a radiographic study. Ultrasonography, computed tomography, voiding cystourethrography and fistulogram can be used. Among these examinations, ultrasonography and computed tomography are reported as the best diagnostic tools.[19] In our study, ultrasonography showed 61.1% diagnostic accuracy, and computed tomography showed 57.1% diagnostic accuracy. Ultrasonography was usually performed first, and if not definitive, computed tomography was subsequently performed. Seventeen of the 21 patients with urachal anomaly could be diagnosed by these two radiographic studies. One patient with urachal cyst could not be diagnosed by radiographic studies, and his diagnosis was confirmed after the operation. Prompted by symptoms such as umbilical discharge and urine leakage from the umbilicus, the remaining three patients (2 with patent urachus and 1 with urachal sinus) were diagnosed purely through surgical exploration.

Rich et al. reported that urachal anomalies were associated with other genitourinary conditions such as hypospadia and crossed renal ectopia.[4,20] Other investigators reported associated anomalies in cases of persistent urachal remnants including meatal stenosis, hypospadias, umbilical and inguinal hernias, cryptorchidism, anal atresia, omphalocele, ureteropelvic obstruction and most frequently, vesicoureteral reflux.[21,22]

Infection has been reported as the most common complication in urachal anomalies.[23] Severe infection may develop into peritonitis and sepsis. In our cases, samples such as umbilical discharge from nine patients were cultured, and Staphylococcal species were isolated in two patients. Peptostreptococcus, Streptococcus and Escherichia coli were isolated in the other three patients, respectively. Adenocarcinoma of the urachal remnant is rare, accounting for less than 0.01% of all malignancies.[24] Other cancers, such as transitional cell carcinoma have also been reported.[25]

Treatment includes surgical repair and antibiotics.[11] In this study, all patients eventually underwent excision and three patients were supplemented by partial cystectomy. After the operation, none of the patients had postoperative complications or any other symptoms. In our opinion, surgery is the optimal treatment modality since urachal remnants do not regress spontaneously.

Our study has some limitations. We investigated symptomatic patients, but asymptomatic urachal anomalies have been reported in some of the literature. To evaluate all urachal anomalies, both symptomatic and asymptomatic patients need to be considered.

In conclusion, our results indicate that the optimal diagnostic radiographic studies are ultrasonography and computed tomography. Ultrasonography showed higher diagnostic accuracy than computed tomography, and the latter has the additional disadvantage of exposure to radiation. For these reasons, we recommend ultrasonography as the first diagnostic tool in the pediatric age group. If this does not produce definitive results, computed tomography may be used as the second modality. Our treatment outcomes also support surgical repair as the optimal treatment since urachal anomalies usually do not disappear spontaneously and can develop into other diseases related to infection or cancer.

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