

Urachal abnormalities: clinical and imaging features

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ABSTRACT

Introduction: The clinical manifestation of urachal abnormalities may mimic many intraabdominal or pelvic diseases. We present clinical, imaging and pathological findings of a spectrum of complicated urachal abnormalities and determine whether imaging can be used to differentiate tumour from infection.

Methods: From January 1993 to December 2006, seven patients with surgically-proven complicated urachal abnormalities had their clinical, imaging and pathological features reviewed.

Results: There were three men and four women, aged 12–73 years. Four patients had infected urachal remnants and three had urachal carcinoma. The main clinical findings in infected urachal remnants were dysuria, abdominal pain and mass. The patients of urachal carcinoma presented with abdominal mass and haematuria. Computed tomography (CT) was performed in all cases, and ultrasonography (US) was performed in four cases. CT in all cases showed a mass located extraperitoneally in the midline just beneath the rectus abdominis muscle and extending from the umbilicus to the dome of the urinary bladder. There were one well-defined cystic mass and six ill-defined solid masses. US showed one cystic mass and three echogenic masses. Cystography was performed in one patient and it showed indentation to the dome of the urinary bladder with mucosal irregularity. The cystic mass and one ill-defined solid mass were pathologically-proven to be xanthogranulomatous inflammation. The other five solid masses were found to be adenocarcinoma in three and chronic non-specific inflammation in two cases.

Conclusion: Preoperative diagnosis of urachal abnormalities may be suggested by clinical presentation and imaging features. However, it is difficult to differentiate tumour from infection based on imaging features alone.

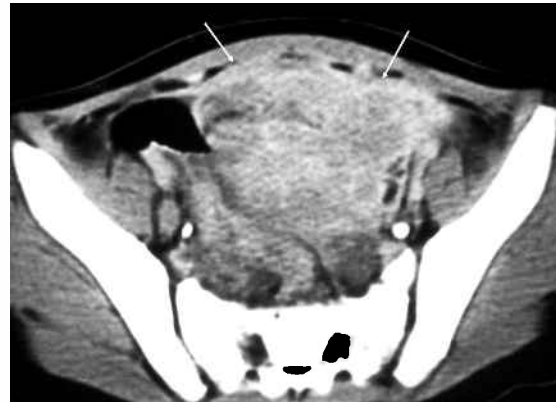


Fig. 1 Xanthogranulomatous inflammation. Contrast-enhanced CT image shows a midline, ill-defined heterogeneously-enhancing mass (arrows) extending from the umbilicus to the dome of the bladder.

Keywords: infected urachal remnants, urachal abnormalities, urachal carcinoma, urachus

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INTRODUCTION

Urachal abnormalities are usually seen in children and are rare in adults.^(1,2) The clinical manifestations of urachal abnormalities may mimic many intraabdominal or pelvic disease processes.⁽²⁻⁴⁾ In the past, the definite diagnosis of urachal abnormalities was frequently made at the time of surgery. Understanding the anatomy and imaging features of urachal abnormalities is important for correct diagnosis and treatment. We present a spectrum of urachal abnormalities, including infection and carcinoma, preoperative diagnosis by ultrasonography (US) and computed tomography (CT) with a review of the developmental anatomy, clinical presentation and imaging features.

METHODS

We retrospectively reviewed the clinical presentation, imaging and pathological findings in four patients with infected urachal remnants and three patients with urachal carcinoma from January 1993 to December 2006. There were three men and four women, aged 12–73 years (mean 43 years). Cystography was performed in one patient. US was performed in four patients and CT was performed in

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Table I. Clinical, imaging and pathological findings of seven cases with urachal abnormalities.

Case no.	Age (years) / gender	Symptoms and signs	Imaging features	Pathology
1	12 / F	Fever, dysuria, abdominal pain	US: mixed echogenic mass with intravesical extension CT: ill-defined heterogeneous enhancing mass	Xanthogranulomatous inflammation
2	20 / F	Abdominal pain, mass	US: ill-defined echogenic mass CT: ill-defined heterogeneous mass	Chronic non-specific inflammation
3	57 / F	Abdominal pain, constipation	CT: ill-defined heterogeneous mass	Chronic non-specific inflammation
4	38 / M	Abdominal mass	US: cystic mass CT: well-defined cystic mass	Xanthogranulomatous inflammation
5	57 / M	Haematuria, dysuria, abdominal mass	US: heterogeneous echogenic mass Cystogram: indentation at the dome of the urinary bladder with mucosal irregularity CT: ill-defined heterogeneous enhancing mass	Adenocarcinoma
6	73 / F	Haematuria	CT: ill-defined, homogenous enhancing exophytic mass at the anterosuperior aspect of the urinary bladder	Adenocarcinoma
7	49 / M	Abdominal mass	CT: ill-defined, heterogeneous enhancing mass with anterior abdominal wall infiltration	Adenocarcinoma

all patients. Diagnosis was confirmed by surgery in all patients.

RESULTS

The clinical, imaging and pathological features of these seven patients with urachal abnormalities are summarised in Table I.

Case 1

A 12-year-old girl presented with fever, dysuria and lower abdominal pain for two weeks. Physical examination revealed mild suprapubic tenderness. The white blood cell count was 19,600/mm³. Urinalysis was normal. US showed a mixed echogenic mass in the anterosuperior portion of the bladder with apparent intravesical extension. CT demonstrated a midline, ill-defined heterogeneously-enhancing mass extending from the umbilicus to the dome of the bladder (Fig. 1). At surgery, the mass was adherent to the sigmoid colon and terminal ileum. Excision of the mass with a 10-cm long ileal resection, and a 10-cm long sigmoidectomy and partial cystectomy were performed. Histopathological examination revealed chronic xanthogranulomatous inflammation with fibrosis. The patient was discharged without any complication.

Case 2

A 20-year-old woman presented with lower abdominal pain and mass. Physical examination showed a 10-cm ill-defined firm mass at the suprapubic region. The full blood count and urinalysis were normal. US revealed an

ill-defined echogenic mass anterosuperior to the bladder with indentation to the dome of the bladder (Fig. 2a). CT showed an ill-defined heterogeneously-enhancing mass deep to the rectus abdominis muscles, extending from the umbilicus to the lower pelvis, with compression of the dome of the bladder (Fig. 2b). The patient underwent surgical exploration, and an irregular firm mass with omental adhesions was found. Wide excision of the mass with partial cystectomy was performed. Pathological examination revealed chronic non-specific inflammation.

Case 3

A 57-year-old-woman suffered from abdominal pain for one month and constipation for several months. Physical examination revealed an ill-defined mass below the umbilicus. The white blood cell count was 10,000/mm³. Urinalysis was normal. CT showed an ill-defined heterogeneously-enhancing mass at the mid-anterior abdominal wall extending from the umbilicus to the dome of the bladder (Fig. 3). The mass was adherent to the transverse colon. Surgical exploration demonstrated a firm mass beneath the rectus abdominis muscle, extending from the umbilicus to the dome of the bladder. The mass was adherent to the transverse colon, sigmoid colon and terminal ileum. En bloc resection of the mass including the umbilicus, a contiguous segment of transverse colon, sigmoid colon, terminal ileum and a cuff of bladder, was performed. An abscess cavity was found in the mass and histological examination revealed chronic non-specific inflammation with fibrosis. She was well postoperation.

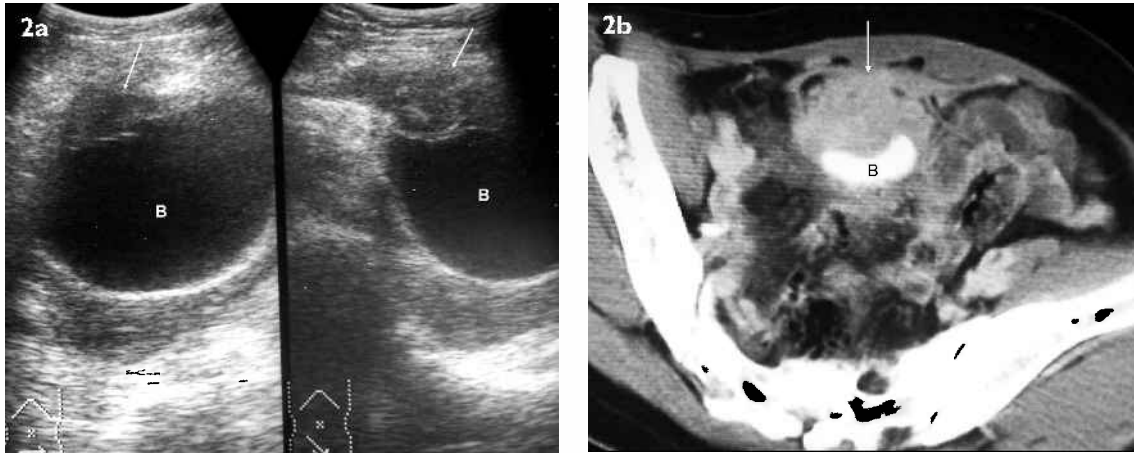


Fig. 2 Chronic non-specific inflammation. (a) Composite US images (transverse [left] and oblique [right]) show an ill-defined mass (arrows) anterosuperior to the bladder (B), with indentation to the dome of the bladder. (b) Contrast-enhanced CT image shows a heterogeneously-enhancing mass (arrow) deep to the rectus abdominis muscle with compression of the dome of the bladder (B).

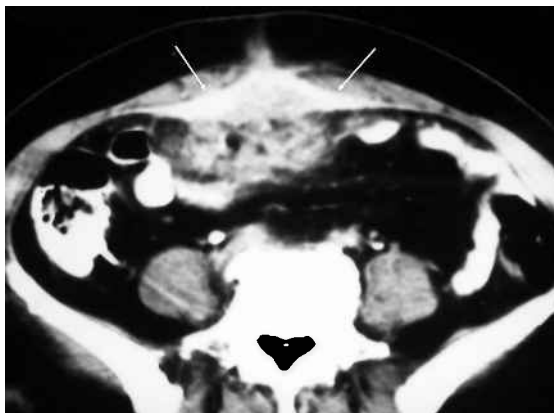


Fig. 3 Chronic non-specific inflammation. Contrast-enhanced CT image shows an ill-defined heterogeneously-enhancing mass (arrows) in the mid-anterior abdominal wall.

Case 4

A 38-year-old man presented with an abdominal mass for one month. Physical examination showed a large tense cystic mass in the lower abdomen. The white blood cell count was $14,600/\text{mm}^3$. Urinalysis showed 4–8 white blood cells and 1–2 red blood cells per high power field. US revealed a large cystic mass in the lower abdomen (Fig. 4a). CT showed a large well-defined cystic mass with a slightly thick-walled enhancement extending from the umbilicus to the lower pelvis, that was adherent to the dome of the bladder (Fig. 4b). At surgery, a 14-cm cystic mass was removed with partial cystectomy. Histopathology revealed xanthogranulomatous inflammation. The postoperative course was uneventful.

Case 5

A 57-year-old man presented with a one-month history of haematuria and dysuria. He had history of an appendectomy four months ago. Physical examination showed an ill-defined tender mass at the suprapubic region. The white blood cell count was $17,700/\text{mm}^3$.

Urinalysis revealed numerous white blood cells and red blood cells. He was considered to have intraabdominal abscess. US showed a heterogeneous echogenic mass in the pelvic cavity anterosuperior to the urinary bladder. Cystogram showed a mass indenting the superior aspect of the bladder with mucosal irregularity. CT demonstrated a midline ill-defined heterogeneously-enhancing mass beneath the rectus abdominis muscle extending from the umbilicus to the dome of the bladder (Fig. 5). Cystoscopy revealed an irregular mass at the dome of the bladder. Biopsy was initially thought to be transitional cell carcinoma. The mass was excised en bloc together with the umbilicus and sigmoid colon, and a radical cystectomy was performed. Histopathological examination showed an adenocarcinoma with bladder and colon invasion. Retrospective review of the previous biopsy confirmed adenocarcinoma. The postoperative course was complicated with enterocutaneous fistula and tumour recurrence.

Case 6

A 73-year-old woman presented with painless haematuria for one day. Physical examination was normal. The full blood count was normal. Urinalysis showed numerous red blood cells and 15–20 white blood cells per high power field. Cystoscopy revealed a 1-cm sessile mass at the anterior wall of the urinary bladder. Biopsy revealed well-differentiated adenocarcinoma. CT demonstrated a midline, ill-defined, homogeneously-enhancing exophytic mass at the anterosuperior aspect of the urinary bladder (Fig. 6). Wide excision of the mass with partial cystectomy was performed. Pathological examination revealed moderately-differentiated adenocarcinoma. The surgical margin was positive for adenocarcinoma. Pelvic irradiation was subsequently performed.

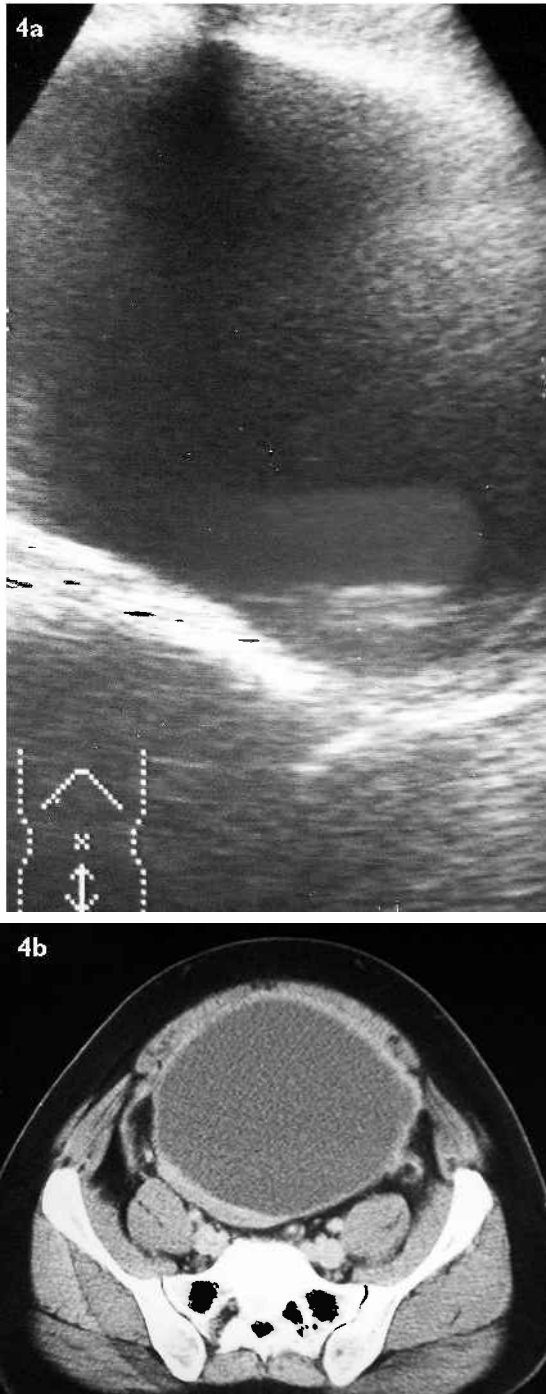


Fig. 4 Xanthogranulomatous inflammation. (a) Sagittal US image shows a large cystic mass in the lower abdomen. (b) Contrast-enhanced CT image shows a large well-defined cystic mass with a slightly thick-walled enhancement extending from the umbilicus to the lower pelvis adherent to the dome of the bladder.

Case 7

A 49-year-old man presented with chronic ulcer and discharge from the umbilicus for one year. Physical examination showed an ulcerative mass at the umbilicus. The full blood count was normal. Urinalysis showed numerous red and white blood cells. Cystoscopy revealed a 1-cm sessile mass at the dome of the urinary



Fig. 5 Adenocarcinoma. (a,b) Contrast-enhanced CT images (5b) obtained at the level lower than (5a) show a midline heterogeneously-enhancing mass (M) beneath the rectus abdominis muscle extending from the umbilicus to the dome of the bladder (B).

bladder. Biopsy from the ulcerative umbilical mass revealed adenocarcinoma. CT demonstrated a large ill-defined heterogeneously-enhancing mass extending from umbilicus to the dome of the urinary bladder with adjacent anterior abdominal wall infiltration (Fig. 7). En bloc resection of a mass and anterior abdominal wall, with partial cystectomy were performed. Histopathology showed moderately differentiated adenocarcinoma. Postoperation course was uneventful.

DISCUSSION

The urachus is an embryological remnant of the allantois. During foetal development, the bladder is continuous with the allantois. Between the fourth and fifth months of development, the urachus narrows to become a fibromuscular strand extending from the apex of the bladder and the umbilicus. The urachus lies in the extraperitoneal space of Retzius between the transversalis fascia anteriorly and the peritoneum posteriorly.^(3,5,6) Histologically, the urachus is composed

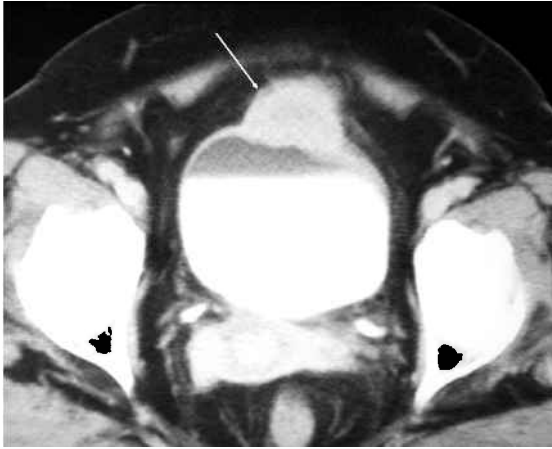


Fig. 6 Adenocarcinoma. Contrast-enhanced CT image shows a midline, ill-defined, homogeneously-enhancing exophytic mass (arrow) at the anterosuperior aspect of the urinary bladder.



Fig. 7 Adenocarcinoma. Contrast-enhanced CT image shows a large ulcerated, ill-defined heterogeneously-enhancing mass extending from the umbilicus (arrow) to the dome of the urinary bladder with adjacent anterior abdominal wall infiltration.

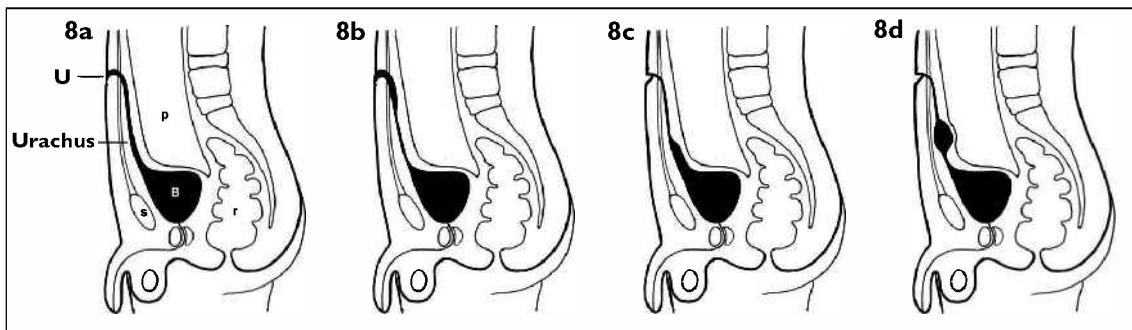


Fig. 8 Diagram shows the four types of congenital urachal anomalies (modified from ref. 3). (a) patent urachus or urachal fistula, (b) umbilical-urachal sinus, (c) vesicourachal diverticulum, and (d) urachal cyst. B: bladder; p: peritoneal cavity; r: rectum; s: symphysis pubis; u: umbilicus

of all three bladder layers, the innermost layer being lined with transitional epithelium in 70% of cases and with columnar epithelium in 30%, a middle submucosal layer of connective tissue, and the outermost muscular layer in continuum with the detrusor muscle.⁽³⁾ Involution of the urachus is usually complete at birth. Incomplete involution results in urachal abnormalities. There are four main types of persistent urachal remnant:^(2,3) (a) patent urachus or urachal fistula, (b) umbilical-urachal sinus, (c) vesicourachal diverticulum, and (d) urachal cyst (Fig. 8).

The patent urachus or urachal fistula usually presents in the neonatal period with a urinary discharge from the umbilicus. An umbilical-urachal sinus, vesicourachal diverticulum or urachal cyst may close normally after birth, but then reopen in association with pathological conditions that are often categorised as acquired disease.⁽³⁾ Acquired urachal disorders most commonly present in adulthood and infection is the most common complication.^(1,3,6,7) The route of infection may be lymphatic, haematogenous or vesical. The clinical symptoms of urachal abnormalities vary depending on the types and complications. They are usually misdiagnosed

as appendicitis, Meckel's diverticulitis, acute prostatitis, urinary tract infection, pelvic inflammatory disease and bladder carcinoma.^(4,8) Diagnostic evaluation of suspected urachal disorders include intravenous pyelography, cystography, sinography, cystoscopy, US and CT.^(1,3-11) Because the urachus is located in the anterior abdominal wall and is not interfered with intestinal structures, CT and US are suited for demonstrating urachal anomalies. A midline supravescical soft tissue mass on US and CT is suggestive of urachal abnormalities. Complex echogenicity at US and heterogeneous attenuation with variable contrast enhancement in and around the disease process at CT make it difficult to differentiate an infected urachal remnant from urachal carcinoma.⁽³⁾ However, a supravescical mass with calcification is highly suggestive of urachal carcinoma because 50%–70% of urachal carcinoma contains calcification.⁽³⁾ Magnetic resonance imaging offers the advantage of multiplanar imaging and may be useful to clearly determine the involvement of the urinary bladder or of other adjacent structures.^(11,12)

Of our seven patients, four were complicated with infection and three had adenocarcinoma. The

main clinical presentations in the four patients with infection were dysuria, abdominal pain and mass while the three patients with adenocarcinoma presented with abdominal mass and haematuria. Preoperative diagnosis of urachal abnormalities could be made in all patients, but differentiation between infection and carcinoma is difficult because both infection and carcinoma can present as a solid mass with adjacent organ involvement. Although the presence of calcification is suggestive of carcinoma, it was not found in our patients. We think that the urachal cyst appears echogenic on US and enhancement on CT because of superimposed infection. The rising white blood cell count may be helpful to differentiate infection from tumour in some cases. Correct preoperative diagnosis of infection could be made only in a case with cystic mass. Of the four cases with infected urachus, two were found to be xanthogranulomatous infection. Xanthogranulomatous lesions are the unusual forms of chronic inflammatory process. Histopathology is characterised by the presence of large lipid-laden macrophages.^(13,14) The urinary tract involvement is most commonly observed as xanthogranulomatous pyelonephritis.⁽¹⁴⁾ Few cases of urachal xanthogranulomatous lesions have previously been reported.⁽¹³⁻¹⁵⁾ Preoperative differentiation from urachal carcinoma was also impossible in these reports.

Malignant urachal neoplasms are also rare, representing less than 0.5% of all bladder cancers.⁽³⁾ 85%–90% of urachal carcinomas are adenocarcinomas, though most of the normal urachus is lined by transitional cell epithelium. This is probably due to metaplasia of the urachal mucosa into columnar epithelium followed by malignant transformation. Less common urachal carcinomas include squamous cell carcinoma, transitional cell carcinoma and sarcoma.^(3,11) Benign urachal neoplasm have been rarely reported.⁽¹⁶⁾ Most of the patients with urachal carcinoma are 40–70 years of age, with males being affected more often than females.^(3,11) Although urachal carcinoma is rare, correct preoperative diagnosis is important for proper management and can be suggested from the clinical history of haematuria, location of the lesion and imaging findings. Ashley et al suggested that the strongest predictors of malignancy in urachal mass were haematuria and age older than 55 years.^(17,18) Treatment of urachal abnormalities usually includes complete excision of the urachus, umbilicus and a cuff of bladder.^(4,6) Total removal of the cyst wall is essential because there is a 30% reinfection rate and carcinoma may develop in an unresected or incompletely resected urachal remnant.^(2,5) The prognosis in cases of urachal carcinoma is worse than

that of primary bladder carcinoma because the tumour arises outside the bladder and involves the mucosa late in the disease process. Local invasion frequently presents before diagnosis.^(11,17)

In conclusion, we report seven cases of urachal abnormalities comprising four infections and three cases of carcinoma. Dysuria, abdominal pain and palpable mass were the main clinical symptoms in cases of infection, while abdominal mass and haematuria were found in carcinoma. The lower abdominal midline mass extending from the umbilicus to the dome of the urinary bladder should guide the diagnosis of a urachal abnormality. US and CT can accurately define these lesions. However, differentiation between infection and carcinoma is difficult on the basis of imaging alone.

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