Urachal cyst with xanthogranulomatous cystitis: A rare case report

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INTRODUCTION

The urachus is the embryological remnant of the allantois, which connects the apex of the bladder to the umbilicus. It usually obliterates fully to become the median umbilical ligament. Urachal defects are uncommon, and cysts are usually asymptomatic. It is an embryonic connection between the urinary bladder and the allantois. It obliterates during early infancy, to form the median umbilical ligament between the transverse fascia and the peritoneum. Histologically, the inner layer is modified transitional epithelium, the middle is fibroconnective tissue, and the outer layer is a smooth muscle layer.1 Xanthogranulomatous changes have been reported to occur in many sites including the colon, ovary, pancreas, salivary gland, appendix, gallbladder, endometrium, brain, and kidney. Here, we present a rare case of urachal cyst with xanthogranulomatous cystitis (XC) in a 30-year-old female who manifested with dysuria and hypogastric mass.

SUBJECTS AND METHODS

A 30-year-old female presented with dysuria and lower abdominal pain. On examination, a hard hypogastric mass was present fixed to the rectus muscle. Routine hematological and biochemical investigations were unremarkable. Urine routine was normal, and culture was sterile. Ultrasound abdomen showed thickening with cystic lesion in the urinary bladder.

Abstract

An urachal cyst is a sinus remaining from the allantois during embryogenesis which is rarely manifested in adults. The urachus is an embryologic remnant which degenerates after the birth. Defective obliteration of the urachus leads to urachal abnormalities. Urachal cyst is a rare pathology in adult women, and this pathology should be considered in the differential diagnosis of acute abdomen. Xanthogranulomatous cystitis (XC) is a benign disease of unknown etiology. The clinical manifestations of these are nonspecific such as lower abdominal pain, umbilical discharge with occasional hematuria. Urachal lesions present with persistent umbilical drainage in infants and newborn. However, in 35% cases, enclosed urachal cyst or infected urachal cyst (abscess) manifests without having umbilical discharge. Computed tomography scan and magnetic resonance imaging are of little help to the identification of these preoperatively. Here, we present a rare case of urachal cyst with XC in 30-year-old female which has produced diagnostic dilemma.

Keywords: Urachal cyst, urachus, xanthogranulomatous cystitis
Contrast-enhanced computed tomography (CT) of the abdomen showed partially distended urinary bladder. A focal well-defined hypodense, noncommunicating cystic lesion was seen measuring 38 cm × 25 cm × 25 cm. It appeared to involve the anterior and superior wall of urinary bladder at dome region [Figure 1a]. Diffuse perilesional mesenteric/omental inflammatory fat stranding area seen anterior and superior to the urinary bladder. Lesion shows single central mildly enhancing internal septations. On this investigation, a diagnosis of urachal carcinoma was suspected. On cystoscopy, edema of anterior wall and dome of urinary bladder was seen. It was followed by biopsy which revealed the features of chronic cystitis. In view of the radiological investigation which revealed a mass, it was decided to perform partial cystectomy. Intraoperatively, the mass was found to be arising from the entire wall of the bladder and was infiltrating into the posterior rectus sheath and muscle. On gross examination, an irregular lesion seen over the dome of the bladder along with attached urachus with umbilicus. Cut surface of lesion showed a cyst measuring 3 cm × 2 cm filled with cheesy material in the bladder [Figure 1b]. Bladder mucosa appears unremarkable. Microscopy revealed persistent urachus attached to the bladder wall forming a cyst lined by mixed inflammatory cell infiltrate [Figure 2a] comprising foamy histiocytes [Figure 2c], plasma cells, lymphocytes, neutrophils [Figure 2b], foreign body, and Langhans giant cells [Figure 2d] are seen. Also noted are granulomas comprising aggregates of epithelioid histiocytes. This inflammation is involving the full thickness of bladder. Overlying bladder mucosa was unremarkable with focal squamous metaplasia. Perivesical tissue at places shows dense inflammation forming germinal center with creeping of fat in the bladder muscles. Umbilicus was unremarkable. No evidence of malignancy was seen. Hence, final diagnosis of persistent urachus with xanthogranulomatous inflammation along with cystic change involving dense bladder, and transmural cystitis change was made. The postoperative course was uneventful; the patient was advised to have antibiotics and he responded well as he came for regular follow-up.

DISCUSSION

The urachus is an embryonic connection between the bladder dome and the umbilicus, which elongates as the bladder descends. The urachus is obliterated by the 5th–7th month of gestation and it forms the median umbilical ligament as a fibrous cord which lies between the transversalis fascia and parietal peritoneum.[1,2] Urachal anomalies are rare in adulthood and are caused by the incomplete obliteration of the urachus. The congenital anomalies of the urachus are patent urachus, urachal sinus, vesicourachal diverticulum, and urachal cyst. Urachal cysts form when both the umbilical and vesical ends of the urachus close while an intervening portion remains patent. Urachal cysts usually become symptomatic when these are infected. Infected urachal cysts present with fever, abdominal pain, and abdominal tenderness with erythema, lower abdominal mass, nausea, vomiting, and dysuria.[3] The diagnosis of urachal cysts is mainly clinic, and the diagnosis is usually confirmed by ultrasonography, CT, and also magnetic resonance imaging. These imaging methods also give information about the size of cyst and its relationship with peripheral tissue. Ultrasound imaging commonly reveals a tubular mass in the midline below the umbilicus.[4] Although staphylococcal species are usually isolated from the culture of abscess, microorganisms such as Escherichia coli. The recommended treatment of the urachal abscess is intravenous antibiotic therapy and total surgical excision. The resection of the cyst wall entirely is
especially recommended. Because of the high recurrence rate and the risk of malignancy, drainage of the abscess is not recommended. Traditionally, surgical excision is performed through laparotomy, but laparoscopic excision is also acceptable. Clinical presentation of the urachal abscess may mimic an acute abdomen. Thus, in the differential diagnosis, other causes of acute abdomen should be primarily considered. Because of urinary symptoms, cystitis and pyelonephritis are also getting involved in differential diagnosis. Although infected urachal cyst is uncommon in adult women, it should be considered in the differential diagnosis of an acute abdomen, especially with a mass in the midline. It should be underlined that infected urachal cysts can be misdiagnosed, especially as acute appendicitis.

XC is a benign chronic inflammatory disease which mostly is associated with urachal diverticula. The disease only presents with lower abdominal pain with cystitis-like symptoms, umbilical discharge, and occasional hematuria. The etiology of XC is not known. A number of theories show about its origin as immunological disorders, abnormal lipid metabolism, metaplasia of urothelium due to chronic infection.

Medical treatment is seen ineffective in many cases, so conservative management is rarely employed. Hence, the choice of treatment is surgical resection. Localized disease can be treated by simple tumor excision. However, when disease is seen with urachal remnant or adenoma, partial cystectomy is employed. In addition, antibiotic therapy and urinary astringents may be helpful. Isolated excision of XC lesion may not be indicated.

CONCLUSIONS

Urachal anomalies are seen rarely in adults, and surgical intervention is required due to rare chance of recurrence and malignancy. As in our case, it is one of the probable pathologies causing diagnostic and treatment difficulties to general surgeons when presents as intra-abdominal mass. Its diagnosis is possible with surgical exploration and histopathological evaluation like our patient.

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Conflicts of interest
There are no conflicts of interest.

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