CASE REPORT

Infected urachal cyst in an adult male: a rare entity

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Abstract:
The urachus is an embryo-logic remnant which in adults attaches the bladder dome to the umbilicus and degenerates after the birth and presents as the midline umbilical ligament. Defective obliteration of the urachus ends up in urachal abnormalities. Urachal cyst is an uncommon clinical condition that is typically symptomless. It is one amongst a spectrum of urachal abnormalities most commonly found in children. They are seldom seen in adults. It is diagnosed via ultra-sonography and computed tomography (CT scan). However, in some cases, the diagnosis is made by means of surgical exploration and histo-pathological analysis.
Case report:
We report a case of infected urachal cyst on a male patient aged 23 years who had a complaint of swelling over umbilicus with transparent fluid leakage for 20 days. There was no previous history of any surgical intervention. Pre-operative evaluation included ultrasound. Patient underwent laparotomy, excision of the tract along with the cuff of the bladder and umbilicus was done.

Keywords: Urachus, infected urachal cyst, laparotomy, transparent fluid leakage

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Introduction:
The urachus is a fibrous remnant of the allantois. It is a canal that drains the urinary bladder of the fetus and runs within the umbilical cord.[1] Incomplete regression of the fetal urachus results in urachal abnormalities. They are more common in children in comparison to adults, as urachus obliterates in early infancy[2] to form the median umbilical ligament between the transverse fascia and the peritoneum. Histo-logically, the inner layer is modified transitional epithelium, the middle is fibro-connective tissue, and the outer layer is a smooth muscle layer.[3] If urachus does not obliterate, urachal anomalies may appear including a patent urachus (50% cases), an urachal cyst (20–30% cases), urachal sinus (10%) and less commonly a vesico-urachal diverticulum.[4,7] In adults, urachal cyst (UC) is the commonest variety, with contamination being the standard method of introduction. Diagnosis remains challenging due to the rarity of this lesion and the non-particular nature of its symptomatology.[2] An infected urachal cyst is one of a spectrum of presentations of urachal pathology, which are all uncommon in adult-hood. Patients tend to present in a hetero-geneous manner, making diagnosis troublesome. Ultrasound (USS), computed tomography (CT) and magnetic resonance imaging (MRI) will help in diagnosis. Deferral in treatment may have serious consequences as complications include sepsis, fistula formation, and rupture leading to peritonitis.[5] The recommended treatment of choice is surgical excision due to the potential risk of malignant change.[4]

Case Presentation:
A young male of 23 years, un-married, with no known co-morbids, presented with swelling over the umbilicus for last 20 days along with transparent fluid leakage from it.

On per-abdominal examination there was a 2x2cm mass over the umbilicus. It was erythem-

atous with irregular margins .On palpation it was firm, non-tender with leakage of clear fluid from it on compression. 2cm peri-umbilical area was indurated and mildly tender. Rest of abdomen was soft and non-tender.

Pre-operative investigation includes ultrasound whole abdomen and pelvis which showed 2.2x3cm heterogeneous mass in anterior abdominal wall not extending intra-peritoneally.

Exploratory Laparotomy was planned and done. Post-operatively there was a fibrous tract extending from umbilicus to the anterior dome of bladder. It was adherent to the anterior abdominal wall and sigmoid meso-colon. Tract was excised along with cuff of bladder and umbilicus. The bladder was repaired. Patient was catheterized with 16 Fr Foley's catheter which was retained for 5 days.

Recovery was unremarkable. He was discharged with medications on 7th post-operative day, and was advised to do follow up in out-patient department. He was followed post-operatively for a month in the outpatient clinic of surgery. He developed no post-surgical complications. Histo-pathological analysis of the resected specimen showed urothelial lining with chronic inflammation with no evidence of malignancy.

Discussion:

The urachus is an embryonic remnant of the cloaca and the allantois which is present after birth as a midline fibrous cord extending from the anterior dome of the urinary bladder to the umbilicus.[6] The cloaca, in the fetal life, is an extension of the urogenital sinus and allantois, and is derived from the yolk sac.[9] After the descent of bladder into the pelvis, the apical part narrows to form epithelialized fibromuscular remnant, the median umbilical ligament or the urachus which lies between the transversalis fascia and parietal peritoneum.[7] Urachal anomalies are uncommon in adult-hood and are caused by the deficient obliteration of the urachus.[8] The incidence of a urachal remnant is 1:300,000 in infants and 1:5000 in adults. This lesion typically affects young adults 20–40 years of age, with a male-female ratio of 2:1.[1] Those remnants found in newborns younger than 6 months usually resolve spontaneously without the need for surgery. Those found in older patients require treatment because of a greater risk for infection, and most importantly, due to an increased risk of malignant transformation.[9]

Incomplete regression of the urachal lumen results in several anomalies:

Patent urachus: A persistent communication between bladder lumen and umbilicus, leading to urine leakage from the umbilicus. Diagnosis can be made by fistulography or cystography.

Urachal cyst: The urachus remains patent between two end points with no connection to the bladder or umbilicus, as shown in our case.

Urachal sinus: The urachus is patent at its umbilical side, but not the bladder, resulting in umbilical mass or inflammation with or without periodic discharge with fever and leukocytosis. A thickened midline tubular structure below the umbilicus can be seen by ultrasound scan (USS).

Urachal diverticulum: The urachus is patent at the bladder dome. Ultra sonography can reveal a non-communicating extra-luminal fluid-filled structure by the umbilicus. Computerized tomography (CT) demonstrating a midline cystic structure at antero-superior aspect of the urinary bladder.[5]

Other than patent urachus, which presents with urinary discharge from umbilicus in neo-natal period, a large portion of anomalies are a-symptomatic, unless complicated by infection. Routes of infection may be hematogenous, direct spread from bladder or lymphatic.[7] A urachal cyst occurs in 1 in 5,000 live-births, but is only clinically relevant in 1 in 150,000 of the population.[11] The incidence of a urachal cyst in adults is unknown yet is uncommon. It is more common in males than females. The most common presentation of urachal abnormality is the urachal cyst, with infection being the typical manifestation. Commonly cultured micro-organisms from the cys-

tic fluid include Escherichia coli, Enterococcus faecium, Klebsiella pneumonia, Proteus, Streptococcus viridans and Fusobacterium.

Clinical signs and symptoms are non-specific and might solely be known once infection happens. The presence of the triad of symptoms including a tender midline infra-umbilical mass, umbilical discharge and sepsis ought to raise the suspicion of urachal cyst infection.[10] Owing to the low incidence and hetero-geneous presentation patients are often mis-diagnosed.[5] If left untreated, it gradually enlarges and may drain through the umbilicus as was seen in our patient, or drain into the bladder or both.[2] Infected urachal cyst will often result in urachal abscess and might develop general septic condition. With a severe urachal cyst infection, pyo-urachus can form a fistula with bladder, intestine or omphalos. Abscess rupture due to enlargement of infected urachal cyst will cause acute abdomen requiring emergency operation.[6] The differential diagnosis of an umbilical mass should include hematoma, abscess, umbilical hernia, urachal carcinoma, tumors of the abdominal wall,[9] vitelline duct anomalies, appendicitis, granulomatous inflammations, and granulation tissue from the umbilical stump.[12]

The diagnosis of urachal cysts is mainly clinic and is usually confirmed by ultra-sonography, computed tomography (CT), and also magnetic resonance imaging (MRI). These imaging ways conjointly offer information concerning the dimensions of cyst and its relationship with peripheral tissue.[8] Cystography or cystoscopy is often helpful to outline the complete extent of the cyst wall and delineate the urachal sinus.[6] Ultrasound scan will facilitate to form diagnosis in 77% of patients.[2] CT is the most vital within the diagnostic work–up. It reveals the kind of urachal anomaly with adequate degree of accuracy[9] however, both infected urachal cysts and urachal carcinomas usually show increased echogenicity at US and thick-walled cystic or mixed attenuation at CT, making it troublesome to differentiate between them.[11] Diagnosis is often made following exploratory laparotomy for an unexplained acute abdomen. Complications of infection include sepsis, fistula formation, and rupture leading to peritonitis, Necrotizing fasciitis has additionally been reported as a rare complication of an infected urachal cyst.[5] Other reported complications include uracho-colonic fistula, stone formation and neoplastic transformation.[2]

Surgical treatment by complete primary excision is the treatment of choice for urachal remnants because of the danger of recurrent cyst infection(30%) and malignant transformation.[1,2,5,10,11] If urachal pathology appears with signs of infection, a two–stage treatment is recommended: initially, administration of antibiotics and resolution of inflammation, followed by surgical removal is the treatment of choice. In benign urachal anomalies, complete excision, with or without the cuff of the bladder, is sufficient. It is not necessary to remove the umbilicus.[9] Because of the high recurrence rate and risk of developing malignancy within the urachal remnant, it is a key point to complete resection of the cyst wall through-out its length during operation. Open excision has been performed as the treatment of choice, traditionally. However, recently, the laparoscopic technique has been accepted as an alternative choice as a result of quicker recovery, less operative pain and better cosmetic results.[6]

Conclusion:
Urachal remnants are a rare condition in adulthood, the correct diagnosis of which is fundamental to the correct treatment. The patient's history and physical examination are crucial for the correct diagnosis. Although now-a-days several methods are available for diagnosing pathology of urachal remnants, none of these is completely accurate. Radical excision of urachal remnants employing a laparoscopic approach is a safe and effective alternative to open surgery. The two-staged operation is the better procedure for achieving a shorter hospitalization and to prevent complications. Laparoscopic excision is the ideal approach nowadays, although the optimal treatment method is still indefinite.

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Dr. Hajrah Hilal Ahmed, collected the data, references and wrote the menuscript

Prof. Dr. M. Jamaluddin, critically review the article and made final changes

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