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Patent Urachus

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Introduction

Patent urachus refers to one condition in a rare spectrum of disorders referred to as urachal anomalies. These conditions result from the failure of the involution of normal embryologic tissues that serve to empty the fetal bladder. The location and amount of persistent tissue dictate the presenting symptoms. Some of these urachal anomalies are obvious at birth, while others are more subtle and not diagnosed until adulthood or only incidentally discovered after imaging is obtained for other reasons. Historically, surgical resection of urachal anomalies was routinely undertaken, given the potential for malignancy in retained ectopic tissue.[1][2][3] Early surgical resection has been challenged by many studies demonstrating not only the low incidence of the condition in general but the even lower chance of future malignancy.[4][5] Accurate and expeditious diagnosis of urachal anomalies in the child with umbilical wetness, persistent drainage, recurrent urinary tract infections, and recurrent umbilical infections is required to assist with symptom relief, avoid recurrent severe infection, and to provide definitive treatment.[5]

Etiology

During the fourth week of embryogenesis, the three-layered embryonic disk folds into a cylindrical shape.[6] The umbilical vessels (two umbilical arteries and one umbilical vein), urachus, and omphalomesenteric duct enter into the now narrowed interface between the yolk sac and embryo. The omphalomesenteric duct connects the developing gut to the yolk sac. The allantois forms to become the urachus, which connects the developing bladder to the umbilicus. Obliteration of the urachus is believed to occur at the end of gestation, with some believing that it occurs in the first few days following birth.[7] After involution, a fibrous cord remains in the preperitoneal space between the umbilicus and the bladder.[1] Failure of involution results in a spectrum of urachal anomalies related to the amount and location of residual tissue.[7] Total failure results in a patent, tubular connection between the urinary bladder and umbilicus. A bladder diverticulum occurs when there is persistent tissue at the bladder with no connection to the umbilicus. Persistent tissue at the umbilicus with no connection to the bladder is called an umbilical polyp or sinus. A urachal cyst results when there is patency along the midportion of the urachus with the closure of the tract at the umbilicus and bladder.

Epidemiology

The incidence of urachal anomaly is considered difficult to determine, as some have urachal anomalies that are asymptomatic or undiagnosed.[7] True patent urachus is believed to be a rare entity, which was confirmed in a review study conducted at the Hospital for Sick Children in Toronto, Canada. Researchers examined the incidence of radiographically diagnosed urachal anomalies in children undergoing abdominal imaging over twelve years.[1] The prevalence of all urachal anomalies in their general pediatric population was 1.03%. True patent urachus was a rare diagnosis, representing only 1.5% of all diagnosed urachal anomalies. The median age of diagnosis varies widely between studies. In general, true patent urachus is diagnosed earlier in life, given the obvious physical exam findings, including near-constant leakage of urine from the umbilicus. Urachal anomalies are associated with posterior urethral

valves, as a patent urachus may be the only relief valve for an otherwise obstructed urinary tract. Consequently, the incidence in male children is three times higher than in female children.[8]

Pathophysiology

The manifestations of a urachal anomaly depend on the location of the remnant tissue.[7][1][2] A true '*patent urachus*' results in a communication from the urinary bladder to the umbilicus, resulting in drainage of urine at the umbilicus, resultant dermatitis, and the potential for recurrent urinary tract infections. The presence of persistent tissue at the umbilicus without connection to the bladder can result in persistent umbilical drainage, dermatitis, and possible umbilical infection. A *bladder diverticulum* results when persistent tissue is connected to the bladder. A bladder diverticulum is typically of no consequence except in the instance when the diverticulum causes ureteral obstruction. A *urachal cyst* results when there is patency along the midportion of the urachus with closure of the tract to both the umbilicus and bladder. Infection of the cyst can occur and result in abdominal or suprapubic pain with a palpable mass.

Histopathology

A noted, but rare, complication of urachal anomalies is malignant degeneration.[2] In the pathologic specimens of half of the adults undergoing resection of urachal anomalies, adenocarcinoma was found. This has never been reported in children undergoing resection. Pathologic examination of urachal carcinomas have demonstrated mucinous, enteric, and signet ring cell types.[9]

History and Physical

The average umbilical cord is 1.5 cm in diameter and 3.6 cm in circumference.[10][11] An abnormally thick cord should prompt further investigation for the potential for urachal anomaly.[12][13][14] If no visible abnormalities are present at birth, some infants and older children are brought to their pediatrician with the complaint of persistent umbilical drainage with classic urinous discharge.[7][1] Umbilical polyps can manifest as umbilical drainage with abnormal appearing tissue at the umbilicus unresponsive to silver nitrate. Other children, particularly those with urachal cysts, present later in life with infection resulting in periumbilical cellulitis, abdominal pain, suprapubic pain, and/or palpable mass.[15] Physical exam should focus on identifying any umbilical defects, sinus tracts near the umbilicus, palpable masses posterior to or just distal to the umbilicus, and examining the umbilicus for movement with micturition or discharge with gentle pressure applied to the bladder.[16][17]

Evaluation

In a newborn with visible umbilical cord abnormality, an ultrasound helps to delineate the diagnosis further.[8][18] If ultrasound demonstrates thickened tissue with a possible connection to the bladder, a repeat ultrasound should be performed in six to twelve months. In the case of an infant with patent urachus, a voiding cystourethrogram should be performed to evaluate for other urinary tract abnormalities, such as posterior urethral valves. In the case of posterior urethral valves, a patent urachus may represent the only pathway for the flow of obstructed urine. In the child with the complaint of persistent umbilical wetness, the physical exam should focus on identifying any abnormal opening in the umbilicus or for any palpable masses posterior or just distal to the umbilicus. If there is a question if the fluid being expressed from the umbilicus is urine, it can be sent for creatinine. If there is an obvious opening, a sinogram can be performed, which involves injecting contrast into the tract.[19] This is typically difficult to perform in children and is more useful in adult patients. Recurrent umbilical or urinary tract infections usually indicate a urachal cyst. The causative organisms in repeat urinary tract infections are often skin flora or gram-negative Enterobacteriaceae. While not usually necessary for diagnosis, CT scans are commonly performed before evaluation by a pediatric general surgeon and are highly sensitive for diagnosis.[20]

Treatment / Management

Patent urachus should be surgically resected, either by an open or laparoscopic approach, to avoid recurrent urinary tract infections and umbilical skin breakdown.[21] In newborns or small children, an open approach typically involves

smaller incisions, less scarring, and is relatively easy from a technical standpoint. The patent urachus should be resected in its entirety, including the portion of the bladder it attaches to, and the bladder should be closed in two layers with absorbable suture. While not mandatory, rarely some surgeons will place a Foley catheter and leave it in place for 24 to 72 hours after surgery, particularly if a large bladder repair is required.[22] In a larger child, laparoscopic repair might be more advantageous, especially if it can avoid a formal laparotomy.[23] Similar recommendations for two-layer bladder repair still apply. If a child is diagnosed with a urachal anomaly and has an infected umbilicus, incision and drainage should be performed, antibiotics started, and the wound should be fully healed for 4 to 6 weeks before the elective repair is considered to avoid a bladder leak. If the child is asymptomatic from the standpoint of a urachal anomaly, particularly with a bladder diverticulum, the risk to benefit ratio of excision has not been determined, and many surgeons are now opting to follow these patients.[7][1][24][4][5] It has even been suggested that low-grade inflammation can obliterate the remnant structure. If resection is not undertaken, the family must be counseled on the future risk of malignancy; however, a study found the number of urachal anomalies required to treat (number needed to treat) to prevent one case of adenocarcinoma was 5,721.[1]

Differential Diagnosis

When a large umbilical cord is observed at delivery, one should consider the potential for the containment of bowel, omphalomesenteric duct, patent urachus, or vascular anomaly.[12][13] An ultrasound should be performed to delineate the cause further, particularly if urachal anomaly (or patent omphalomesenteric duct) is suspected.[25] The complaint of persistent moistness of the umbilicus in an infant is normally from an umbilical granuloma, which is typically observed as a red, raw-appearing heap of tissue in the umbilicus. This can be treated in the office with silver nitrate.[26] Failure to respond to silver nitrate can indicate the presence of an umbilical polyp, which can be omphalomesenteric or urachal in origin.[27][28] Patent omphalomesenteric duct can also cause drainage from the umbilicus, but its appearance is consistent with enteric contents. While urachal cyst can present as a palpable umbilical mass or umbilical swelling, this overwhelmingly represents an umbilical hernia in children. The diagnosis of omphalitis in a newborn cannot be overlooked in the case of purulent umbilical drainage with surrounding induration, erythema, and tenderness.[29]

Prognosis

The prognosis of children with isolated urachal anomalies is excellent however, the potential presence of posterior urethral valves should not be overlooked in infants with total patent urachus. It should also be noted that 25% to 30% of children with prune belly syndrome have patent urachus.[30][31][32]

Complications

With regards to patent urachus, recurrent urinary and umbilical infections can represent a significant cause of morbidity. While postoperative complications are rare, wound infection and dreaded intraperitoneal urinary leak can occur.[21]

Postoperative and Rehabilitation Care

Some pediatric general surgeons place Foley catheters before every patent urachus repair, while others forgo this step.[22] Baths and total submersion of surgical incisions in water should be avoided for the first two weeks postoperatively. A routine postoperative visit is conducted to ensure incision(s) are healing well. Children can generally be trusted to self-regulate their activity, and the risk of subsequent incisional hernia is rare.

Consultations

Consultation with a pediatric surgeon is required in patients with a suspected or confirmed urachal anomaly. In children with associated urinary tract abnormalities (i.e., posterior urethral valves), urology is routinely involved.

Deterrence and Patient Education

Families should be educated on the etiology of urachal anomalies. If diagnosed in infancy, the potential for other urinary defects (like posterior urethral valves) and the need for further workup should be discussed. The families of infants brought in for evaluation of umbilical wetness should be reassured that this likely represents self-limiting umbilical granulation tissue. In children with a clear diagnosis of patent urachus, previously infected urachal cyst, or symptomatic urachal polyp or diverticulum, surgical resection and the expected recovery should be thoroughly reviewed. In children with incidentally discovered urachal anomalies for whom conservative management is being considered, the rare potential for the future development of cancer should be made clear.[4][5]

Pearls and Other Issues

Diagnosis of urachal anomalies can be elusive, but persistent umbilical drainage should raise the concern for a urachal anomaly. While this condition is typically found in isolation, the potential for posterior urethral valves in an infant with patent urachus should not be overlooked.[8] Voiding cystourethrogram should be utilized in these infants for further evaluation of the urinary tract. In addition, evaluation for patent urachus should be undertaken in children with prune belly syndrome, as it is found in nearly one-third of children with the syndrome.[30][31][32] This diagnosis should also not be missed in the differential diagnosis of a child with an abscess near the umbilicus. In the case of an infected urachal cyst, treatment of the infection is required to decrease tissue inflammation before elective repair several weeks later.[21]

Enhancing Healthcare Team Outcomes

While sometimes apparent at birth, patent urachus and other urachal anomalies may have a periodic, more insidious presentation. Frontline healthcare providers, like primary care providers and emergency department physicians, must the diagnosis of patent urachus or urachal remnant and their different presentations as part of the differential diagnosis of a child who is brought in for persistent umbilical drainage, recurrent urinary tract infections, recurrent umbilical infection, and/or a palpable mass near the umbilicus. When utilizing imaging modalities like ultrasound, a skilled sonographer and radiologist familiar with these diagnoses are critical. A pediatric general surgeon should be involved with any suspect urachal anomaly or when umbilical drainage initially thought to be an umbilical granuloma does not improve with silver nitrate application. Pediatric urology involvement may be necessary when other urologic abnormalities are diagnosed. Untreated urachal anomalies were historically thought to all inevitably degenerate into malignancy; however, this is now recognized as an extremely rare occurrence.[1] [Level 4]

Questions

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