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Case Report

Urachal Anomaly: A Case Report

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Abstract

Urachus is a tubular structure that extends cranially from the anterior dome of the bladder to the umbilicus. From the level of the umbilicus, the bladder progressively descends and reaches the pelvis where the urachus stretches and eventually, the lumen progressively obliterates and is replaced by a fibrous tract in early adult life. Failure of this obliteration can lead to anomalies of the urachus such as patent urachus, urachal sinus, and urachal cyst. Urachal anomalies is a diagnostic dilemma and may be confused with obstructed hernia, appendicitis, Meckel diverticulitis, urinary tract infection, pelvic inflammatory disease, and bladder carcinoma. The cyst remains largely asymptomatic unless infected and may present with acute symptoms such as suprapubic pain, dysuria, fever, nausea, vomiting, haematuria, pelvic pain, and purulent umbilical discharge. Imaging techniques such as ultrasonography, CT, and MRI are the main modalities employed for the diagnosis of a urachal cyst. All urachal anomalies should be surgically removed regardless of whether they are asymptomatic or acutely symptomatic.

Keywords: *Urachus; Urachal Cyst; Urachal Sinus; Urachal Fistula; Urachal Anomalies; Patent Urachus*

Introduction

Urachal anomalies arise from the incomplete obliteration of urachus which is a primitive structure that connects the umbilical cord to the fetal bladder [1]. Urachus is a tubular structure that extends cranially from the anterior dome of the bladder to the umbilicus. The clinical presentation is variable and is evident at birth with umbilical cord abnormalities or diagnosed later in adolescence with complaints of persistent umbilical wetness or recurrent umbilical infection. The incidence of urachal cyst is one in 5,000 live births [2].

Aim of the Study

The aim of the case report is to describe the anatomy and embryology, discuss the different types of congenital Urachal anomalies, outline the appropriate evaluation and enumerate the management options available for urachal anomalies.

Case Report

A 9 year old male child presented with pus discharge from umbilicus of 2 days duration which was profuse, yellowish, non-blood stained and foul smelling. He also complained of abdominal pain which was peri-umbilical, insidious onset, diffuse, non-radiating, mild intensity, continuous with no aggravating or relieving factors. There was no history of fever or bladder, bowel alterations.

The general and systemic examination did not reveal any significant abnormality. On per abdominal examination, there was tenderness in the umbilical region. The umbilicus was midline and inverted with minimal surrounding erythema, induration and minimal seropurulent, foul smelling discharge. There was no external opening and there was no increased local rise in temperature. A 2 x 1 cm globular shaped periumblical swelling with well-defined edges, fluctuant and cystic in consistency was present.

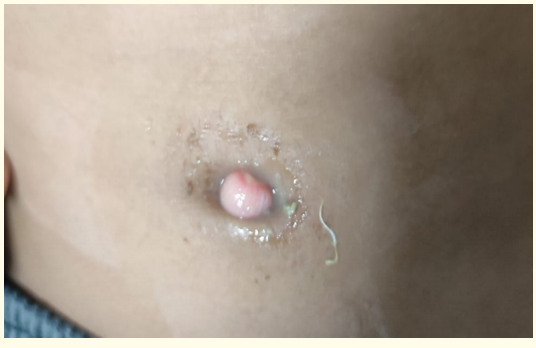

Figure 1: *Umbilical swelling and discharge.*

Imaging

Ultrasonography revealed a heterogeneous collection at the umbilicus measuring 1.5 x 3.1 x 2 cm (Volume - 4.5 cc). There was a thin tubular structure extending from the collection and reaching upto the dome of urinary bladder suggestive of infected urachal sinus with collection.

Contrast enhanced computerised tomography was suggestive of a linear, soft tissue density seen in the expected location of urachus with its proximal and distal ends thickened with a thin mid part. Proximal thickened part measured 1.7 x 1.2 x 2.4 cm, with no opacification of structure filling through the bladder which suggested an of absence of fistulous tract.

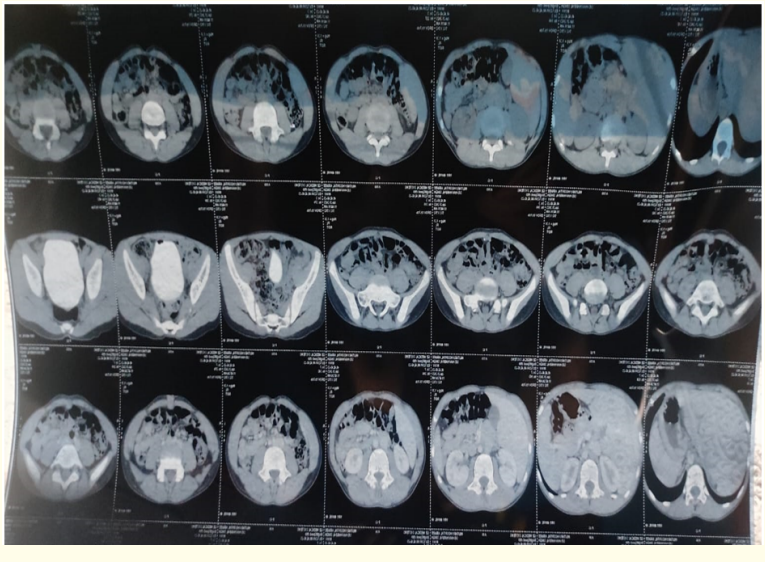

Figure 2: *CT scan image suggestive of a linear, soft tissue density seen in the expected location of urachus with its proximal and distal ends thickened with a thin mid part.*

Management: The patient was managed conservatively with oral antibiotics, analgesics and regular sterile dressings followed by Excision of sinus tract.

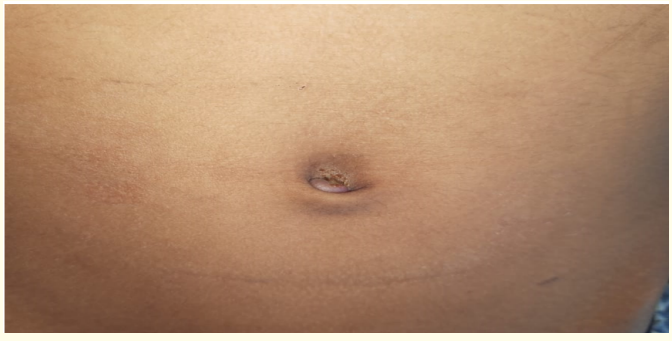

Figure 3: *Post-operative status.*

Discussion

The urachus is a midline structure arising from the anterior aspect of the fetal bladder which in turn is derived from the ventral part of urogenital sinus [1]. The cloaca gives rise to the primitive urogenital sinus and anorectal canal structures between the fourth and seventh week of gestation. Allantois arises from the yolk sac and extends to the cephalic extension of the urogenital sinus, which is the precursor of the fetal bladder. Initial development of the fetal bladder occurs at the level of the umbilicus where the cephalic extension of the urogenital sinus is contiguous with the ventral allantois.

From the level of the umbilicus, the bladder progressively descends and reaches the pelvis by the fourth or fifth month of gestation, where the urachus stretches, and eventually, the lumen obliterated. During the later part of fetal life, the urachus progressively obliterates and is replaced by a fibrous tract in early adult life [2]. Failure of this obliteration can lead to anomalies of the urachus such as patent urachus, urachal sinus, and urachal cyst [2]. The urachal cyst is the most common urachal anomaly and is caused when both the proximal and distal portions of the urachus obliterate leaving a cystic cavity in the middle [3].

Congenital urachal anomalies can be classified into patent urachus, urachal cyst, urachal sinus and vesical diverticulum.

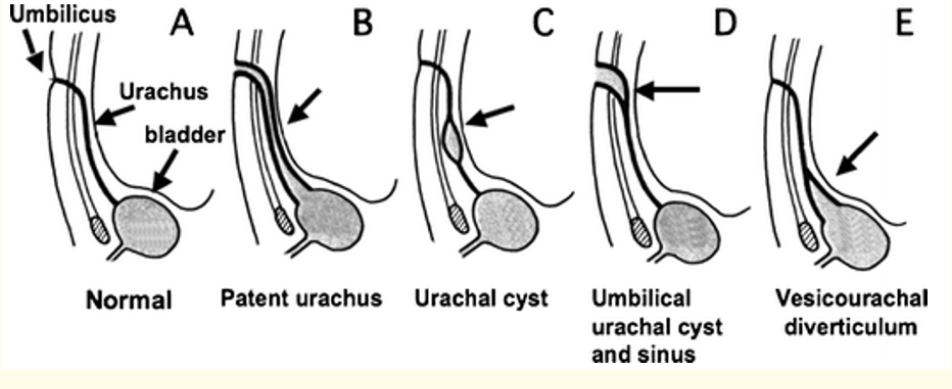

Figure 4: *Classification of urachal anomalies.*

Presentation

The cyst remains largely asymptomatic unless infected [4]. Infected urachal cysts present with acute symptoms such as suprapubic pain, dysuria, fever, nausea, vomiting, haematuria, pelvic pain, and purulent umbilical discharge.

Differential diagnosis

In view of the diverse clinical presentation, the diagnosis of urachal cyst is often confused with obstructed hernia, appendicitis, Meckel diverticulitis, urinary tract infection, pelvic inflammatory disease, and bladder carcinoma [5].

Imaging

Imaging techniques such as ultrasonography, CT, and MRI are the main modalities employed for the diagnosis of a urachal cyst [4]. USG is the most common imaging modality in the diagnosis of patients and attributed due to non-invasive procedure that is readily available in most clinical settings. Fistulography is another diagnostic option useful in observing a sinus at the base of the umbilicus. Its use has been suggested when there is discharge from the umbilicus without the presence of a palpable mass and to differentiate vitelline duct remnants from Urachal remnant. CT scan is a preferred imaging modality that helps secure diagnosis when USG or fistulography are inconclusive or if the patient is acutely ill with signs of systemic infection. MRI has the ability to provide a more detailed picture of the soft tissues and can detect hollow urachal tube, neoplasm, cyst, or abscess. Cystoscopy can be performed which can provide direct visual evidence of urachal diverticula or sinus.

Complications

The delay in diagnosis and treatment of the urachal cyst gives rise to various complications such as peritonitis, uracho-colonic fistula, stone formation, and neoplastic transformations [6].

Management

All urachal anomalies should be surgically removed regardless of whether they are asymptomatic or acutely symptomatic. Removal of a non-infected urachal anomaly will help avoid the possibility of emergency surgery in the future, which undoubtedly carries higher risk and also potential for turning neoplastic. The majority of urachal neoplasms are asymptomatic in the early stages of development, resulting in diagnostic delays and poor prognosis. It is theorized that neglected urachal anomalies might become silent, allowing for repeated exposure to chronic inflammation and infection, increasing the probability of carcinogenesis. Additionally, persistent urachal anomalies have been associated with recurrent UTI and stone formation.

Conservative approach, including antibiotic treatment and regular monitoring of UA is a useful management strategy. Studies utilizing the conservative method have provided evidence that many lesions will spontaneously resolve. Thus, experts assert this method may be particularly useful in incidental findings of UR.

Open surgical excision was the standard protocol for removal of UR, excising the entire urachus and tract, as well as the portion in communication with the bladder dome. Laparoscopic excision of urachal anomalies is now the preferred surgical method. In both adults and children, the laparoscopic approach has demonstrated to be safe, effective, with reduced invasiveness, allows for precise tissue dissection, diminishes blood loss, decreases hospital stay, and faster convalescence and provides far better cosmetic results. Excision of the cyst can be done under antibiotic cover in one sitting or a two-staged procedure can be performed, which involves incision and drainage followed by delayed excision of the urachal remnant [7]. A two stage procedure is an alternative method of surgical management wherein

the first portion of the two-stage procedure includes the surgical drainage, followed by a culture of the lesion's contents. Following drainage, patients are treated with the proper antibiotic therapy. After perioperative drainage, the urachal remnant will be excised entirely. The two-step surgical procedure has been noted to reduce the frequency of postoperative complications.

Conclusion

Urachal anomalies extend cranially from the anterior dome of the bladder to the umbilicus and arise from the incomplete obliteration of urachus which is a primitive structure that connects the umbilical cord to the fetal bladder. Congenital urachal anomalies can be classified into patent urachus, urachal cyst, urachal sinus and vesical diverticulum. Imaging techniques such as ultrasonography, CT, and MRI are the main modalities employed for the diagnosis of a urachal anomalies. The delay in diagnosis and treatment of the urachal cyst gives rise to various complications such as peritonitis, uracho-colonic fistula, stone formation, and neoplastic transformations. All urachal anomalies should be surgically removed regardless of whether they are asymptomatic or acutely symptomatic. Excision of the cyst can be done under antibiotic cover in one sitting or a two-staged procedure can be performed, which involves incision and drainage followed by delayed excision of the urachal remnant.

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