VOLUME - 11, ISSUE - 11, NOVEMBER - 2022 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

General Surgery

MULLERIAN CYST PRESENTING AS A LARGE ABDOMINOPELVIC CYST CAUSING OBSTRUCTIVE UROPATHY IN MALE: A RARE CASE.

Dr. Nayeemunnisa General Surgery Resident, Deccan College Of Medical Sciences, Hyderabad.

Dr. G. Ravi Professor, Deccan College Of Medical Sciences, Hyderabad.

ABSTRACT

Introduction: Mullerian duct cyst presenting as a large abdominopelvic cyst is a rare entity. A Mullerian duct cyst represents an incomplete focal regression of the embryological Mullerian duct in a male. ***Case Report:*** This is a case of a 55 year old gentleman presenting with a pelvic mass of chronic duration with a recent onset of pain abdomen and obstructive urinary symptoms. Radiological imaging revealed a large centrally located cystic pelvic mass and Left Hydroureteronephrosis. The patient underwent Left DJ stenting followed by an Explorative laprotomy and Marsupialization of the cyst. ***Conclusion:*** It is prerogative to understand the development of the male urinary tract and the reproductive system to diagnose rare midline pelvic pathologies and decide on surgical options for treatment of the same. In addition, the final diagnosis being dependent on the histology of such pelvic masses makes it an additional diagnostic challenge for the surgeons.

KEYWORDS : male pelvic cyst, Mullerian duct cyst, large abdominopelvic cysts.

INTRODUCTION

Pelvic cysts presenting as centrally located swellings in a male patient are a rare presentation with clinical symptoms directly proportional to increase in size of the cyst and compression of adnexal structures or organs. Most of them are benign in nature but may cause complications such as urinary tract infections, pain, post voiding incontinence, recurrent epididymitis, prostatitis, hematospermia and infertility [1]. The differential diagnosis for such large centrally located cysts in a male patient include pelvic abscesses, retroperitoneal tumours, Müllerian duct cysts, Hydatid cyst and malignant pelvic tumours [1,2]. Congenital pelvic cysts are a rare entity that may present as a solitary swelling or may be associated with other genitourinary abnormalities such as ipsilateral renal agenesis, hypospadias, and cryptorchidism [2]. The diagnosis of such pelvic cysts is ultimately based on histological confirmation thus making it a challenge for the surgeon. Understanding the embryologic development and normal anatomy of the male distal genitourinary tract can be helpful in evaluating these cysts. The Leydig cells secrete TESTOSTERONE which stimulates the Mesonephric duct/Wolffian duct to form the epididymis, the vas deferens, seminal vesicles, the common ejaculatory duct and the prostate gland. The Sertoli cells of the testis secrete the MULLERIAN INHIBITING FACTOR(MIF)/ANTI MULLERIAN HORMONE (AMH) that inhibits the growth of the Paramesonephric duct/the Mullerian duct, thus preventing the formation of fallopian tubes, the uterus and the upper 2/3 rd of the vagina in a male foetus [3,4]. Persistent Mullerian Duct Syndrome (PMDS) results from alterations present in anti mullerian hormone secretion, gene, or receptors [3,4]. Paramesonephric/Mullerian duct remnants in male include Appendix of Testis (Hydatid of Morgagni) and prostatic utricle. Anomalous absorption of the Mullerian ducts in the male could lead to cystic anomalies in the distal urinogenital tract. Such a phenomenon could account for development of a Mullerian duct cyst or enlarged utricle in adult life. These anomalies could be counterpart to retained vaginal cysts in females [5].

CASE REPORT:

55year old gentleman presented to the emergency department with complains of 8days of sudden onset of severe lower abdominal pain, associated with high grade fever for 5days, constipation and decreased urination since 4days. The patient gave history of presence of a suprapubic mass for the past 4 months. Clinical examination revealed the presence of a 10x15cm soft non mobile tender smooth swelling occupying the hypogastric, umbilical, parts of the RIF and LIF, tender on palpation mimicking a distended urinary bladder [FIGURE 1]. No guarding or rigidity was present. Family history was non contributory. Vitals on preliminary examination were normal. Foleys catheterization was done at the time of admission which resulted in no change of the size of abdomen ruling out acute urinary retention.

***Investigations*:**

Ultrasound of abdomen and pelvis revealed a large 144x108mm cystic lesion with multiple septations and internal echoes on the left side of the pelvis pushing the bladder to right along with left hydroureteronephrosis. A CECT abdomen revealed the same with an additional comment on compression of the distal ureter causing significant upstream dilatation of the left ureter and pelvic calcyceal system [FIGURE 2]. Blood investigations indicated mild anaemia (Hb9.4) and leucocytosis (WBC 24,230) with a serum creatinine value of 2.5 mg/dl. After initial fluid and antibiotic treatment along with pain management, a decision was taken to perform a USG guided aspiration of the pelvic cyst which demonstrated haemorrhagic dark brown fluid with a cytology report suggestive of an inflammatory smear largely comprised of neutrophils and lymphocytes [FIGURE 3]. Preoperatively performed Echinococcus IgG ELISA test was negative.

The provisional diagnosis was taken as Hydatid cyst of the urinary bladder versus congenital distal urogenital cyst and a preoperative consent was taken for Left DJ stenting followed by Explorative laprotomy.

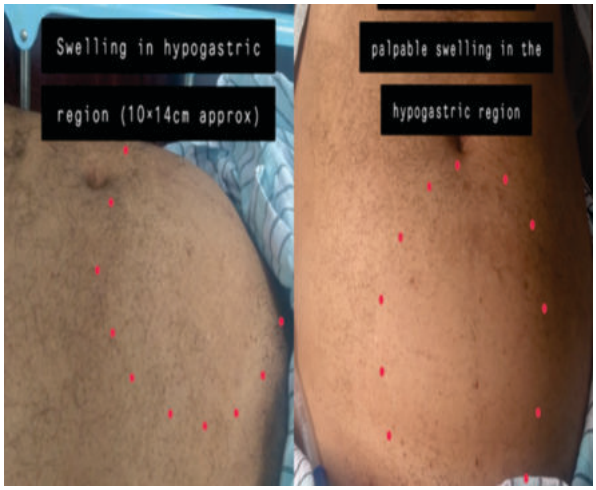

Figure 1 Clinical examination showing approximately 10x14cm suprapubic tender non mobile mass.

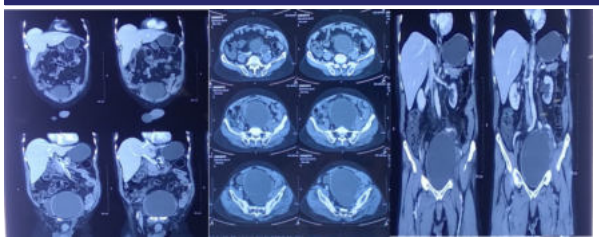

Figure 2 CECT Abdomen showing a large cystic lesion pushing the bladder to the right causing left hydroureteronephrosis

Figure 3 Fluid Aspirated From Cyst, Haemorrhagic In Appearance.

***Intraoperative findings*:**

A 10x 15cm thick wall cyst anterior to the bladder pushing the bladder anterolaterally occupying the left pelvic cavity in close proximity to the iliac vessels and displacing the left ureter posteriorly and medially [FIGURE 4]. The cyst was decompressed, the content were inflammatory fluid with flakes in the dependent portion of the cyst and haemorrhagic fluid around 1000ml in quantity [FIGURE 5]. The right wall of the cyst was densely adhered to the bladder thus a decision was made to perform resection of cyst roof and Marsupialization of the cyst [FIGURE 6]. The position of the iliac vessels and ureters were confirmed on table and dissected free [FIGURE 7]. Intraoperative we considered the diagnosis of Hydatid cyst of the bladder versus a congenital cyst of the distal urogenital tract. A pelvic drain was placed and abdomen was closed. The excised anterior wall and fluid contents of the cyst were sent for Histopathological examination. The patient recovered well with no post operative complications. The serum creatinine reverted back to normal postoperatively. The patient was discharged on day 5 post operatively. The fluid cytology revealed acute inflammatory cells with no evidence of spermatozoa. Histopathological examination demonstrated the presence of inflammatory granulation tissue with mixed cell population and wall lined by fibro adipose tissue with no evidence of laminated wall or scolices or atypia. There was no evidence of malignancy. The diagnosis was an infected Mullerian cyst. The patient was followed for a period of 1 year post operatively with uneventful recovery and no evidence of recurrence.

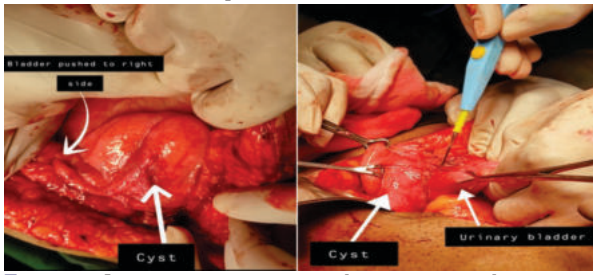

Figure 4 Intraoperative images demonstrating large cyst pushing bladder anterolaterally.

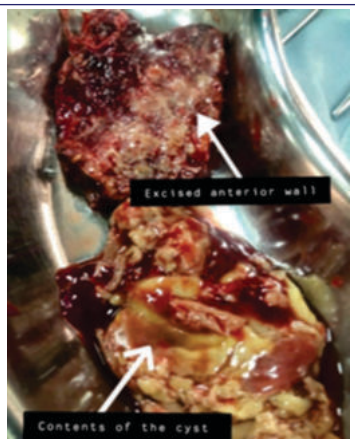

Figure 5 Contents of the cyst along with excised anterior wall.

Figure 6 Marsupialization Of The Cyst; Posterior Wall Of The Cyst

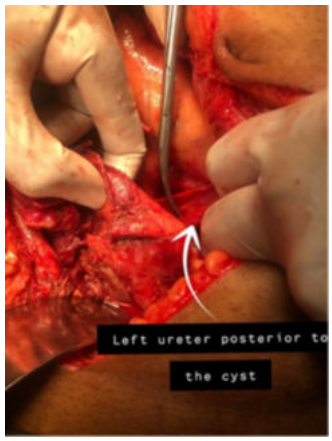

Figure 7 Position Of Left Ureter Confirmed Intra-operatively.

DISCUSSION

Large abdominopelvic cysts of chronic duration in a male patient are rare and thus present a diagnostic challenge. A thorough review of the history, presenting signs, symptoms, clinical examination and laboratory investigations yield valuable information in the preoperative period. Radiological investigations remain standard modalities for confirming the anatomical location of such cysts and their relation to adnexal organs. Transrectal Ultrasound (TRUS) is said to be the most useful modality for detection of such cysts associated with infertility. TRUS is a safe and non-invasive method, so it is highly recommended for assessing the lesions of the prostate, seminal vesicles, and the ejaculatory ducts [6]. Computed tomography is used to depict the exact size, location, and position of the cyst. PELVIC MRI will depict midline cystic lesions that may contain pus/ Hemorrhage if infected. An MRI abdomen and pelvis thus remains the gold standard. Using three-dimensional capability, MRI is able to depict the exact tissues and their contrasts [6]. Based on clinical and

radiological evidence the differential diagnosis of such large chronic midline abdominopelvic cysts in a male patient resulting in obstructive pathology can be narrowed down to Congenital acquired masses like a Mullerian /Utricle cyst , infective aetiologies like Hydatid cyst of the urinary bladder and retroperitoneal tumours or pelvic malignancies.

Infective cysts such as Hydatid cyst of the urinary bladder can be diagnosed based symptoms and critical clinical clues like the endemic nature of the disease. The serological tests like Indirect Haemagglutination test and ELISA for Echinococcal IgG antibodies are useful for the initial screening of suspected cases [7]. Confirmation of test can be done by detecting specific echinococcal antigens. Eosinophilia is present in <25% of infected individuals [7]. Involvement of the urinary bladder is rare. The pathogenesis of bladder hydatid cyst is explained by hematogenous spread. The bladder hydatid cyst clinically remains silent for long periods followed by sudden onset of urinary retention. USG is effective in the diagnosis of hydatid cysts. USG findings in hydatid cysts vary from simple/calcified cyst or cyst with floating membranes (detached endo cyst) also called as "water Lily sign"[8]. Surgical excision and confirmation with histopathological evidence of germinal wall, daughter cysts /scolices remains the mainstay of diagnosis.

Midline abdominopelvic congenital cysts like Mullerian and utricle cyst present in a similar fashion with few critical differentiating features. Mullerian duct cysts are anatomically located above the prostate gland and do not communicate with the urethra with clinically normal genital organs[9]. The age of presentation is usually around the 4th to 5th decade of life [9], commonly as an incidental single pelvic cyst ,rarely as a retroperitoneum cyst[10] .Association with renal agenesis are been reported in literature[11]. Clinical signs and symptoms are directly proportional to the size of the cyst and the presence of infection [12]. Most Müllerian duct cysts are asymptomatic but they may present with irritative urinary symptoms (urinary frequency, urgency), obstructive symptoms (dysuria, decreased urinary flow rate), haematuria, hematospermia, bloody urethral discharge, ejaculatory pain, urinary tract infection, epididymitis, infertility, or constipation [12]. Malignant degeneration is a rare complication[13]. Histologically a variety of features have been described in literature from the presence of mucin secreting tall columnar cells to granulation tissue with mucinous epithelial lining and ciliated columnar cells[12]. Some cyst walls have been reported to have smooth muscle and loose fibrous connective tissue[10].A positive Immunohistochemistry for cytokeratin 7, ER, PgR , oestrogen and progesterone receptors and PAX-8 and immunonegetivity for CK20 has been noted[12,14].

Prostatic utricle cyst is usually seen during the first to second decades of life, typically lies between the bladder and the rectum palpable on per rectal exam. Ten percent to 25% of the cases show an association of renal agenesis/dysgenesis[15] and 25% cases have hypospadias[16].]. Large abdominopelvic utricle cyst is extremely rare and has been reported [15, 16]. Voiding cystourethrogram (VCUG) and retrograde urethrography (RUG) can define the utricle size, which usually ranges from a few millimetres to more than 2 cm. Persistence or untreated prostatic cyst could be a cause of infertility [17]. Morphologically, prostatic utricle cyst appears as a small, single, smooth, unilocular cyst of variable size with urethral connection that may yield semen or blood on aspiration [18]. The cyst lining is cuboidal, columnar, squamous or transitional type. [15]. Malignancy may arise in 3% of the cases; Squamous cell carcinoma is known to occur.

Treatment options for Mullerian cysts include modalities like Trans rectal USG guided aspiration and injection of sclerosing agents [19], however surgery remains the mainstay. Surgical interventions include transurethral cyst incision drainage, Transurethral resection combined with seminal vesiculoscopy for obstruction-associated aspermia [20].Open cyst resection can be performed via transperitoneal, perineal, posterior Para sacral or transvesical transtrigonal route. In large pelvic or abdominal cyst, open surgical excision is the treatment of choice. Laparoscopic cystectomy; Endoscopic treatment has been limited to unroofing infected and obstructed cysts. Recurrences are rare after complete surgical excision but have been reported in a few cases [21].

A follow up period of one year in our patient demonstrated no post operative complications or any evidence of recurrence.

CONCLUSION:

The diagnosis of large abdominopelvic cysts in a male patient presents a clinical challenge. Mullerian duct cysts though congenital in origin can present in adulthood as a large abdominopelvic cyst and should be considered in differential diagnosis of male pelvic cysts. Laboratory investigations and Radiological modalities can help preparing the surgeon in deciding the operative course. Open surgical resection and marsupialization is enough instead of aiming for total excision of the cyst wall with reasonably good outcome.

REFERENCES:

1. Shebel HM, Farg HM, Kolokythas O, El-Diasty T. Cysts of the lower male genitourinary tract: embryologic and anatomic considerations and differential diagnosis. Radiographics. 2013 Jul-Aug;33(4):1125-43. doi: 10.1148/rg.334125129. PMID: 23842975.
2. Pacheco EO, Houat AP, Velloni FG, Blasbalg R, Yamauchi FI, Bittencourt LK, Guimarães CTS. Male Pelvic Cysts: A Didactic Diagnostic Approach. Radiographics. 2021 Oct;41(6):E179-E180. doi: 10.1148/rg.2021210175. PMID: 34597233.
3. Wilson D, Bordoni B. Embryology, Mullerian Ducts (Paramesonephric Ducts) [Updated 2022 Jul 25]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK557727/
4. Mullen RD, Behringer RR. Molecular genetics of Müllerian duct formation, regression and differentiation. Sex Dev. 2014;8(5):281-96. doi: 10.1159/000364935. Epub 2014 Jul 12. PMID: 25033758; PMCID: PMC4378544.
5. Max Maizels. Cambells Urology : Seventh Edition Volume 2, chapter 51, page 1581.
6. Pahlavan F, Taghi Dizaj AV. Similar mullerian cysts in an identical twin: A case report and literature review. J Med Ultrasound [cited 2022 Nov 11]. Available from: http://www.jmuonline.org/preprintarticle.asp?id=335109
7. Arif SH, Mohammed AA. Primary hydatid cyst of the urinary bladder. BMJ Case Rep. 2018 Nov 8;2018:bcr2018226341. doi: 10.1136/bcr-2018-226341. PMID: 30413452; PMCID: PMC6229223.
8. Ganie F A, Dar O H, Kaleem A, Hassan S, Gani M. Hydatid cyst of urinary bladder. Indian J Nephrol 2013;23:462-3
9. Jaidane, M., Hidoussi, A., Slama, A. et al. An acute urinary retention in an old man caused by a giant müllerian duct cyst: a case report. Cases Journal 2, 203 (2009). https://doi.org/10.1186/1757-1626-2-203
10. Tolani MA, Bello A, Ahmed M, Maitama HY. Symptomatic Müllerian cyst in the retroperitoneum of an adult male: Case report and review of the literature. Arch Med Surg 2019;4:53-5
11. Devaraju S, Nerli RB, Hiremath MB. A large mullerian duct cyst presenting as an abdominal mass with ipsilateral renal agenesis: an unusual presentation. Nephrourol Mon. 2012 Fall;4(4):640-3. doi: 10.5812/numonthly.4566. Epub 2012 Sep 24. PMID: 23573507; PMCID: PMC3614304.
12. He J, Tang K. A Müllerian cyst in a male adolescent: a case report and literature review. J Int Med Res. 2021 May;49(5):3000605211016663. doi: 10.1177/03000605211016663. PMID: 34024184; PMCID: PMC8142523.
13. Iura, A., Sasajima, Y., Katsumata, N. et al. Serous adenocarcinoma of the retroperitoneum, as a type of multifocal müllerian carcinoma. Int J Clin Oncol 14, 254–257 (2009). https://doi.org/10.1007/s10147-008-0827-6
14. Naem A, Dlewati A, Alhimyar M, Ousta MA, Alsaid B. A rare presentation and recurrence of a retroperitoneal Müllerian cyst in a male patient: A case report. Int J Surg Case Rep. 2019;65:301-304. doi: 10.1016/j.ijscr.2019.11.001. Epub 2019 Nov 12. PMID: 31760217; PMCID: PMC6883337.
15. Momin YA, Dhende NP, Ghodke BA, Ansari SA, D'Costa GF, Mahajan VR. An abnormally large prostatic utricle cyst associated with unilateral renal agenesis. Urol Ann. 2013 Apr;5(2):129-31. doi: 10.4103/0974-7796.110016. PMID: 23798876; PMCID: PMC3685746.
16. Wang W, Wang Y, Zhu D, Yan P, Dong B, Zhou H. The prostatic utricle cyst with huge calculus and hypospadias: A case report and a review of the literature. Can Urol Assoc J. 2015 May-Jun;9(5-6):E345-8. doi: 10.5489/cuaj.2381. PMID: 26029314; PMCID: PMC4439243.
17. Saito T, Komeya M, Usui K, Kuroda S, Takeshima T, Takashima K, Ikeda M, Kondo Y, Yumura Y. A case of obstructive azoospermia secondary to genitourinary tract infection caused by a prostatic utricle cyst. Asian J Androl. 2022 Sep-Oct;24(5):558-559. doi: 10.4103/aja2021112. PMID: 35042312; PMCID: PMC9491043.
18. Feutry G, De Perrot T, Wirth GJ, Montet X, Martin SP. Prostatic Utricle Cyst as the Most Likely Cause in a Case of Recurrent Episodes of Hematospermia. Case Rep Urol. 2017;2017:7502878. doi: 10.1155/2017/7502878. Epub 2017

Dec 21. PMID: 29430319; PMCID: PMC5752985.
19. Matsui T, Oobe S, Ooka H, Nagata H. [A müllerian duct cyst treated by injection of minocycline: a case report]. Hinyokika Kiyo. 1993 Jan;39(1):69-72. Japanese. PMID: 8460591.
20. Miao C, Liu S, Zhao K, Zhu J, Tian Y, Wang Y, Liu B and Wang Z: Treatment of Mullerian duct cyst by combination of transurethral resection and seminal vesiculoscopy: An initial experience. Exp Ther Med 17: 2194-2198, 2019
21. Gupta GG, Pandey AP. Mullerian duct cyst: Presenting as recurrent abdominal mass. Indian J Urol 2001;17:176-7