Journal of Pediatric Urology (2007) 3, 151—155

Journal of Pediatric urology

CASE REPORT

Ectopic opening of the vas deferens into a Müllerian duct cyst☆

Adriano L. Gomes [a], Luiz G. Freitas Filho [a,b,*], Jovelino Q.S. Leão [a], Antônio C. Heinisch [a], José Carnevale [a]

[a] *Department of Urology, Hospital Infantil Darcy Vargas, São Paulo, Brazil*
[b] *Department of Surgery, Universidade Federal de São Paulo, Escola Paulista de Medicina, Hospital Infantil Darcy Vargas, São Paulo, Brazil*

Received 9 April 2006; accepted 23 May 2006
Available online 10 July 2006

KEYWORDS
Müllerian ducts;
Cysts;
Urinary tract infections

Abstract Two consecutive cases of ectopic opening of the vas deferens into Müllerian duct cysts are presented, with descriptions of the presentation, physical examination, radiographic and ultrasonographic images, surgical findings and treatment undertaken. In both cases, ablation of the cyst and ligature of the vas deferens was the technique employed. Both patients are currently well, with normal urinary flow and no urinary tract infections, 3 years after surgery. Ectopic opening of the vas deferens into Müllerian duct cysts is a rare entity but should always be considered when a child presents with purulent urinary discharge and abdominal tumor. Surgery is the only treatment recommended.

Introduction

The Müllerian duct cyst is an embryo remnant located between the ejaculation ducts that are displaced sideways. It appears as a cystic dilatation of the prostatic utricle, a Müllerian remnant usually present in boys [1—4]. Müllerian duct cysts do not usually develop early in embryological life and seldom present with genital ambiguity [5].

Cystic lesions arising from Müllerian remnants are relatively rare, with more than 100 cases having been reported. The association of Müllerian duct cysts with ectopic opening of the vas deferens into the cyst is an extremely rare congenital alteration, with as few as seven cases reported in the literature by 1992 [6]. Here, we report two

☆ The paper should be credited to Department of Urology, Hospital Infantil Darcy Vargas, São Paulo, Brazil.

* Corresponding author. Department of Surgery, Universidade Federal de São Paulo, Escola Paulista de Medicina, Hospital Infantil Darcy Vargas, São Paulo, Brazil. Tel.: +55 11 5083 5952; fax: +55 11 5083 6690; mobile: +55 11 8426 2658.
E-mail addresses: lfreitasf@terra.com.br, luizfreitasf@hotamail.com (L.G. Freitas Filho).

1477-5131/$30
doi:10.1016/j.jpurol.2006.05.008

cases of children whose deferent ducts ran into large Müllerian cysts.

Case reports

Case 1

VSJ was 1 year and 7 months old, male, with several events of massive pyuria and fever starting from the 6th month of life. He was urinating with normal stream. At physical examination abdomen palpation was normal except for the hypogastric region which was occupied by a painless moving mass of well defined limits. There was purulent discharge from his penis. Urine cytobacteriological tests showed leukocytes $>10^5$, *Escherichia coli* and enterococcus spp. $>10^5$ cfu having been isolated. Ultrasound revealed a thickened bladder wall with normal renal upper tract. VCUG revealed a normal bladder with mild compression of the posterior urethra (Fig. 1). CT revealed a highly increased prostate volume (97 cm^3), with massive cystic formation in its interior (Fig. 2). MRI showed 'expanding' formation with thicker walls and cystic content (Fig. 3). The patient underwent laparotomy under a Pfannenstiel incision that revealed a cystic lesion of the thickened and hardened walls in the retro-vesical position, with the deferens ducts ending in a continuation of the postero-superior wall of the cyst (Fig. 4). The seminal vesicles could not be identified. The cystic lesion was surgically removed and the deferens ducts were tightly tied. The patient was discharged from hospital on day 4 with a vesical catheter that was removed on day 8. Postoperative follow up was uneventful with no urinary flow alterations. Histopathological exam revealed a whitened and elastic tissue membranous specimen measuring 50 × 40 × 30 mm with an unspecific and intense chronic inflammatory process and fibrosis in smooth muscle tissue specimens. Currently, 3 years after the surgical procedure, the patient has a normal urinary flow.

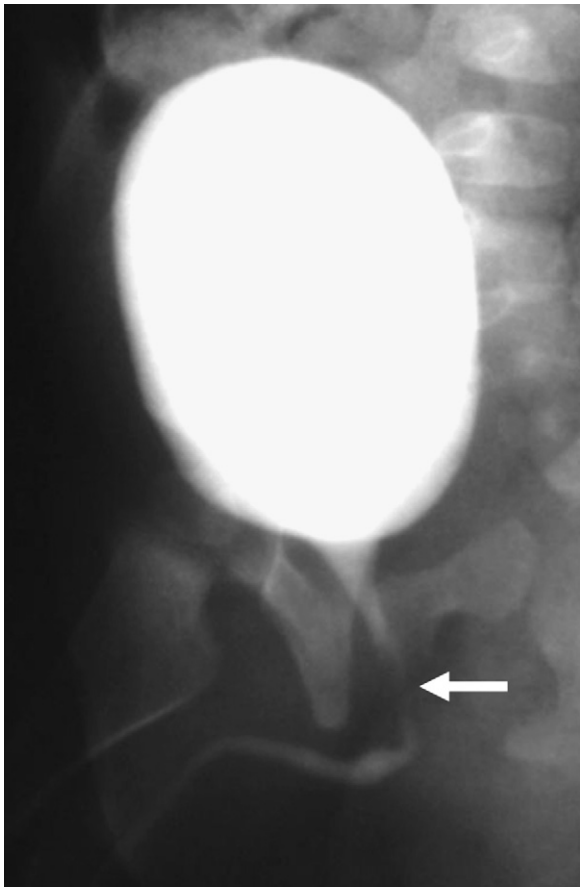

Figure 1 Arrow showing discrete compression of the posterior urethra by the Müllerian cyst.

Case 2

HF was 2 months old and male. Antenatal ultrasonography revealed renal malformation, evolved in the postnatal period, with UTI and purulent discharge in the diaper. He had a palpable mass in the hypogastric area, approaching the umbilicus. Ultrasound at month 1 of life showed a normal left kidney, right kidney with cystic images and a large retrovesical mass (Fig. 5). The patient underwent laparotomy under a Pfannenstiel incision that revealed a thickened wall mass, in the retrovesical position, with the vas deferens ending into the postero-lateral wall of the cyst. The cyst was removed and the vas deferens tightly tied; as in the previous case, the seminal vesicles could not be identified. The vesical catheter was maintained

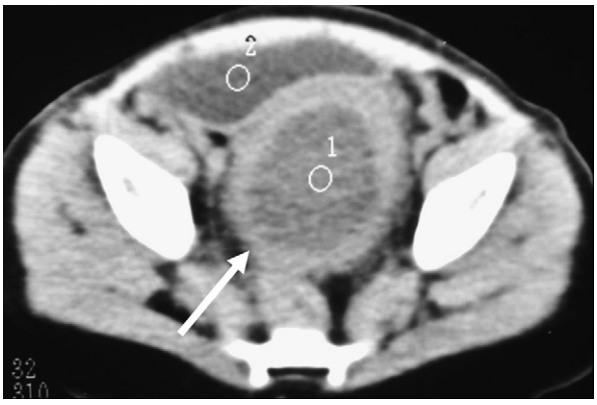

Figure 2 CT scan of the pelvic region. Arrow showing augmentation of the prostate with the Müllerian cyst at its center.

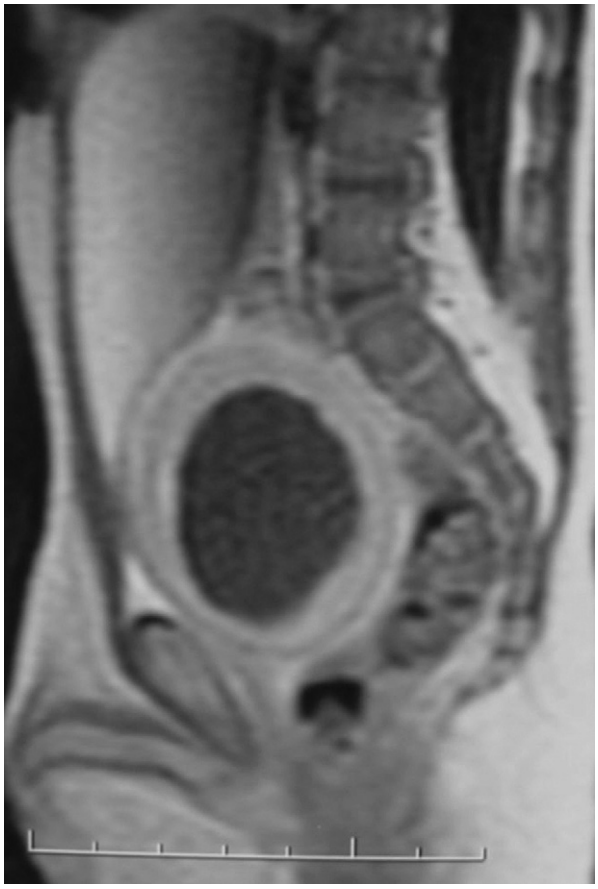

Figure 3 MRI showing the Müllerian cyst.

for 8 days. Histopathological exam of the cyst also revealed an unspecific chronic inflammatory process. Currently, 3 years after the surgical procedure, the patient is well, with a normal urinary flow.

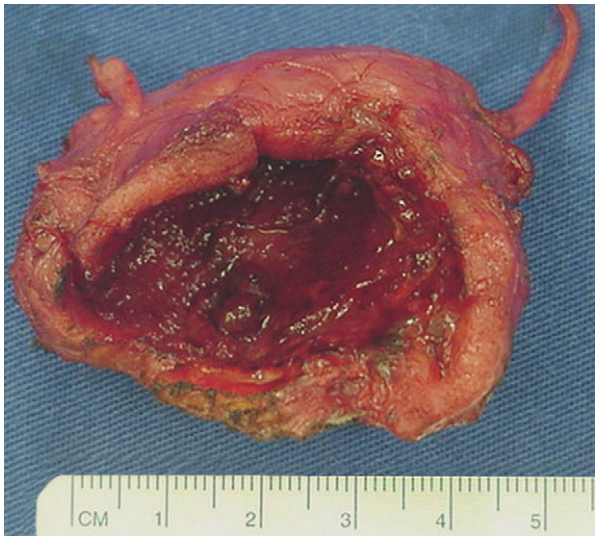

Figure 4 Macroscopic aspect of the ectopic opening of the vas deferens into the Müllerian cyst with agenesis of the seminal vesicles.

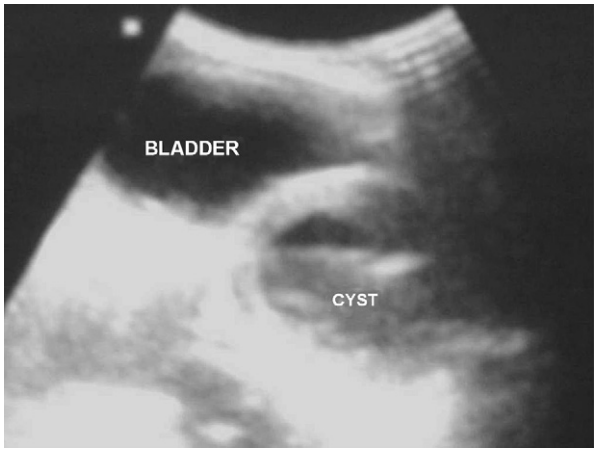

Figure 5 Müllerian cyst compressing the bladder.

Discussion

Until the 30-mm stage, all human fetuses are endowed with two pairs of internal genital ducts: the Wolffian or mesonephric ducts, and the Müllerian or para-mesonephric ducts [7]. So, up to the 8th week of gestation the human reproductive organs show no sign of sex differentiation, and consist of unipotential Wolffian and Müllerian ducts, and bipotential sinusal and external genital primordial organs [7]. The transformation of primitive gonad into testicle and its posterior differentiation are followed by the secretion of a glycoprotein anti-Müllerian hormone, the Müllerian inhibiting substance (MIS), by the Sertoli cells. Müllerian ducts are very sensitive to MIS only during a very short period of development, up to the 8th gestational week in the human fetus [7].

Leydig testicular cells, at the 8th week of gestational age, start producing testosterone. Testosterone, by a paracrine action, transforms the Wolffian ducts, which are initially the excretory ducts of the mesonephros, into epididymis, vas deferens and seminal vesicle. This transformation occurs only when renal function has been taken over by the definitive kidney. Wolffian duct stabilization cannot be obtained by systemic testosterone administration: Wolffian ducts are not maintained in female pseudo-hermaphrodites, even in cases where it can be shown that the virilizing agent reached the fetus very early in pregnancy [7].

The first step of somatic male differentiation is Müllerian duct regression, which is almost always completed at 10 weeks. Male orientation of the urogenital sinus is characterized by prostatic development and the regression of vaginal development. Prostatic buds appear at 10 weeks at the side of the Müllerian tubercle and grow into solid branching cords. Maturation of the prostatic gland

is accompanied by development of the prostatic utricle, the male equivalent of the vagina [7].

Müllerian duct remnants can affect boys and manifest either as a massive prostate utricle, as Müllerian duct cysts, or even as a 'male' vagina that can emerge from the posterior urethra. The persistence of these duct remnants is probably caused by fetal testis incapacity to induce regression of the female structures, as a result of an inappropriate or delayed secretion of MIS [8,9]. Such genitourinary tract anomalies can be found in children with normal external genitalia, but also in those with hypospadias, in particular proximal, or even in children with sexual differentiation anomalies, such as in those with mixed gonadal dysgenesis or true hermaphroditism [3].

The massive augmentation of the prostatic utricle appears as a tubular structure that connects with the urethra, and is frequently found in patients with proximal hypospadias, sexual differentiation disorders, or with other diseases such as prune belly syndrome, Down's syndrome, posterior urethral valves and anorectal diseases [5,8,9]. When other Müllerian structures, such as the uterus, uterine neck and/or uterine tubes are also present, the resulting complex is defined as a 'male vagina'. Another manifestation of Müllerian remnants is in the form of cysts, varying in size from a few centimeters to masses that take over the entire pelvic region. In these cases, the external genitalia are frequently normal, and the patients become symptomatic at an older age [10].

Müllerian duct remnants can manifest clinically through voiding symptoms such as urinary retention and epididymitis, or as a palpable mass at digital rectal examination, or even as an obstructive azoospermia. Symptoms are associated with the size of the cyst, the level of obstruction of the bladder neck, seminal vesicles or ejaculation ducts, or the occurrence of associated infectious processes [11].

Müllerian cysts do not usually connect with the urethra, and thus are rarely identified during a VCUG or a urethroscopy [5]. The VCUG can, in cases where the cyst is too large, show an extrinsic compression of the urethra, or even of the bladder neck (Fig. 2). Ultrasonography reveals a cystic mass posterior to the prostatic urethra and the bladder. CT scanning and MRI can also precisely identify the Müllerian ducts remnants, but in relation to ultrasonography they do not necessarily provide any additional information [1].

In the investigation of cystic lesions of the male pelvis, conducted by Rieser and Griffin [12], four different types are cited: Müllerian duct cysts, ejaculation duct diverticula, seminal vesicle cysts and prostate retention cysts, with Müllerian cysts being the most common. Takahashi et al. [6] cited the median location as the only characteristic that defines the presence of a cyst originating from a Müllerian remnant, but some authors have recognized that the ejaculation duct cysts can also be located in the median position [13–15]. When the seminal vesicles are well developed, the anomaly is most likely more distal and smaller, and the cysts can be treated with an endoscopic incision, enabling relief of symptoms and recovery of fertility in a large number of cases [4,15]. In cases where the vas deferentia end directly into the cyst, with agenesis of the seminal vesicles, as in our two patients, the alteration is more complex, the cysts are characteristically larger, requiring removal, and sterility is inevitable [2].

So far there are no convincing explanations for the absence of the seminal vesicles, as found in our two patients. Glenister [2] showed the utricle to have a dual histological origin: Wolff cells in the more caudal portion and Müller cells in the more cranial portion. Gruenwald observed, in an 8-mm embryo, that the proximity between the Wolff and Müller ducts makes it difficult to say that they may grow independently when a malformation develops [16]. In the embryo observed by Gruenwald, the Wolff and Müller ducts' caudal terminations were so close to one another, with no basal membrane separating their cells, that this could be the most plausible explanation for the variety of pathological findings in Müllerian cyst cases.

The histological characteristics of the cyst wall vary and are of little value for the diagnosis [8]. In our patients, the histological exam revealed an intense, unspecific and chronic inflammatory process and fibrosis in a smooth muscle tissue specimen.

Müllerian cysts must always be treated surgically. The perineal approach must be avoided due to possible infertility [8]. Kaplan et al. [17] used the posterior approach, which involves coccyx amputation, lateral retraction of the rectum and cyst removal. The posterior access, however, is not suitable for large cysts. Transabdominal excision is the usual treatment for obstructive and bulky Müllerian duct cysts. The video laparoscopic approach can be a good alternative for minor lesions [18].

References

[1] Morgan RJ, Williams DI, Pryor JP. Müllerian duct remnants in the male. Br J Urol 1979;51:488–92.

[2] Glenister TW. The development of the utricle and of the so-called ''middle'' or ''median'' lobe of the human prostate. J Anat 1962;96:443–55.

[3] Currarino G. Large prostatic utricles and related structures, urogenital sinus and other forms of urethrovaginal confluence. J Urol 1986;136:1270—9.
[4] Hendry WF, Pryor JP. Müllerian duct (prostatic utricle) cyst: diagnosis and treatment in subfertile males. Br J Urol 1992;69:79—82.
[5] Ritchey ML, Benson Jr RC, Kramer SA, Kelalis PP. Management of Müllerian duct remnants in the male patient. J Urol 1988;140:795—9.
[6] Takahashi M, Kaneko S, Ogawa I, Yamabe K, Tsukada H, Nishida K. A case of ectopic opening of vasa deferentia into Müllerian duct cyst. J Pediatr Surg 1992;27:761—3.
[7] Josso N. The undervirilized male child: endocrine aspects. BJU Int 2004;93:3—5.
[8] Devine Jr CJ, Gonzalez-Serva L, Stecker Jr JF, Devine PC, Horton CE. Utricular configuration in hypospadias and intersex. J Urol 1980;123:407—11.
[9] Shima H, Yabumoto H, Okamoto E, Orestano L, Ikoma F. Testicular function in patients with hypospadias associated with enlarged prostatic utricle. Br J Urol 1992;69:192—5.
[10] Lima M, Aquino A, Domini M, Ruggeri G, Libri M, Cimador M, et al. Laparoscopic removal of Müllerian duct remnants in boys. J Urol 2004;171:364—8.
[11] Desautel MG, Stock J, Hanna MK. Müllerian duct remnants: surgical management and fertility issues. J Urol 1999;162:1008—14.
[12] Rieser C, Griffin TL. Cyst of prostate. J Urol 1964;91:282—6.
[13] Takatera H, Sugao H, Sakurai T. Ejaculatory duct cyst: the case for effective use of transrectal longitudinal ultrasonography. J Urol 1987;137:1241—2.
[14] Goldwasser BZ, Weinerth JL, Carson 3rd CC. Ejaculatory duct obstruction: the case for aggressive diagnosis and treatment. J Urol 1985;134:964—6.
[15] Ng WT, Kong CK, Wong MK. Opening of both vasa into a müllerian duct cyst. J Pediatr Surg 1993;28:756—7.
[16] Gruenwald P. The relation of the growing Müllerian duct to the Wolffian duct and its importance for the genesis of malformations. Anat Rec 1941;81:1—19.
[17] Kaplan GW, Picconi JR, Schuhrke TD. Posterior approach to Müllerian duct and seminal cysts. Birth Defects Orig Artic Ser 1977;13:241—5.
[18] Valla JS, Carfagna L, Tursini S, Mohaidi MA, Bosson N, Steyaert H. Congenital seminal vesicle cyst: prenatal diagnosis and postnatal laparoscopic excision with an attempt to preserve fertility. BJU Int 2003;91:891—2.