AN UNUSUAL AND RARE CAUSE OF RECURRENT EPIDIDYMIS IN AN INFANT

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ABSTRACT
This report describes a rare and unusual case of recurrent epididymitis in a male infant who was found to have a large utricular cyst. Aspects of diagnosis and different methods of treatment are also discussed.

Key words: Utricular cyst, Mullerian duct remnants, recurrent epididymitis.

Introduction
Utricular cyst is a very rare congenital anomaly that arises from incomplete regression of the Mullerian ducts and commonly seen in males with perineal or penoscrotal hypospadias and in intersex disorders (1,2,3,4). Usually they are asymptomatic discovered incidentally but when symptomatic typically they present with urinary tract infection, a palpable abdominal mass, post void dribbling, urethral discharge and rarely recurrent epididymitis (1, 5, 6,7). In this report, we describe a rare and unusual case of utricular cyst in a male infant who presented with recurrent epididymitis outlining aspects of diagnosis and the different modalities of treatment.

Case report
A 1 1/2 year old male child presented to our hospital with a painful left sided scrotal swelling. There was a history of 3 previous similar attacks; the first one was when he was 8 months of age. He was a product of full term normal vaginal delivery and clinically, he had no other abnormalities apart from an enlarged left-sided scrotal swelling that was tender. He had a scrotal ultrasound and a Doppler scan which revealed an enlarged left epididymis and testis with no vascular compromise (Fig. 1). He was admitted to the hospital as a case of left epididymoorchitis and started on antibiotics. He improved, but continued to have an enlarged tender left epididymis. He had an abdominal and pelvic ultrasound which revealed a large cystic swelling in the pelvis measuring 4x3.8x3.2 cm located behind and to the left of the urinary bladder. This was confirmed by CT-scan (Fig. 2). The swelling was in close contact with the urinary bladder. He had a barium enema which was normal. He underwent laparoscopy which revealed a cystic swelling in the pelvis because it was difficult to locate exactly the origin and boundaries of the swelling, it was decided to convert the procedure to open. Prior to this a size 8F catheter was passed into the urinary bladder. Through a Fennisteal incision, and via an extra peritoneal approach, the urinary bladder was mobilized. The urinary catheter accidentally passed into the cystic swelling which was located behind the urinary bladder and in front of the rectum. The nature of the swelling as well as its origin could not be determined, so it was decided to put a suprapubic catheter and close the wound. On the 7th postoperative day, he underwent radiological evalua-

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Figure 1. Scrotal ultrasound showing enlarged left testis and epididymis.
tion using the suprapubic catheter and the catheter in the cystic swelling which revealed a large cystic swelling arising from the prostatic urethra representing a utricular cyst (Fig. 3). One month later, he underwent reexploration. The left vas was found entering the upper part of the cystic swelling while the right one was found adherent to the wall of the cyst and it was difficult to be sure that it was not entering the cyst. The possibility of subsequent fertility was discussed with the family preoperatively and they were not willing for total excision, so it was decided to do near total excision of the cyst taking the left vas with the swelling and preserving the right one. Postoperatively, he did well and was discharged home in a good general condition. Now, he is 6 months postoperatively doing well with no recurrence of his epididymitis and his left epididymis feels normal. Histology of the resected swelling revealed fibrocollagenous tissue with smooth muscle fibers from the wall of the cyst which is lined in areas by squamous epithelium and in other areas by granulation tissue.

**Discussion**

Embryologically, in the male fetus the Mullerian ducts regress under the influence of anti-Mullerian hormone (Mullerian inhibiting substance) (8, 9). This is a glycoprotein secreted by the Sertoli cells of the fetal testes at 8 gestational weeks (8, 9). Persistence of the Mullerian ducts as result of failure of synthesis or action of Mullerian inhibitory substance results in persistent Mullerian duct syndrome (8). This is characterized by the presence of uterine tissue and fallopian tubes in a phenotypic and genotypic male. Utricular cysts are thought to result from incomplete regression of the Mullerian ducts or incomplete androgen-mediated closure of the urogenital sinus caused by an error in the production or sensitivity to local testosterone or anti-Mullerian hormone (1, 10). These cysts are differentiated anatomically from Mullerian duct cysts. Utricular cysts communicate with the urethra and have a tubular or vesicular shape and the majority is seen in younger patients in association with proximal hypospadias and various intersex disorders (1, 4, 10, 11). In contrast to utricular cysts, Mullerian duct cysts present later in life and are associated with normal external genitalia. Mullerian duct cysts generally do not communicate with the urethra and so can not be visualized with a micturating cystourethrogram or a retrograde urethrogram (1, 4, 10, 11). Based on this, it has been postulated that the Mullerian duct cysts are the remnants of the paramesonephric ducts (12). Our patient is unusual because all the features are in favor of a utricular cyst that presented in the neonatal period and communicated with the urethra but at the same time our patient had normal external genitalia.

The presentation of utricular cysts is variable, but many are asymptomatic, and therefore, they escape detection. Symptomatic utricular cysts may present with a variety of symptoms including recurrent urinary tract infection, urethral discharge, post void urine dribbling, urinary retention, urinary incontinence and rarely epididymitis (1, 5, 6, 7). Our patient had recurrent epididymitis that started in early infancy. We recommend that
in a male infant with recurrent epididymitis, utricular cyst should be included in the differential diagnosis. These patients should be investigated with this in mind including abdominal and pelvic ultrasound and CT scan as well as a micturating cystourethrogram and or an ascending urethrogram. Ikoma et al in 1985 classified utricular cysts into four types depending on the size of the cyst and the site of communication with the urethra (4). Our patient belongs to type III.

The definitive treatment of symptomatic utricular cyst is surgical excision but the approach remains controversial. Several surgical approaches have been described including endoscopic transurethral cyst catheterization and aspiration, cyst orifice dilatation, incision and deroofing, transperineal cyst aspiration and sclerotherapy, electrofulguration, open excision (perineal, suprapubic extravesical, transperitoneal, parasacral, transvesical, transstrigonal, retropubic, posterior sagittal with rectal retraction, transrectal posterior or anterior sagittal approach) and recently laparoscopic excision (1, 2, 3, 4). We adopted a suprapubic extravesical and extraperitoneal approach and found this a relatively easy and direct approach. Such an approach was however reported to have a high rate of incomplete excision (13). It is feared that total excision may compromise fertility by traumatically damaging or accidentally excising both vas deference because of their close proximity or more commonly insertion in the utricular cyst. This was the case in our patient where the left vas was terminating within the cyst and had to be sacrificed. The right vas was running within the wall of the cyst and in order to preserve it, we had to do a near total excision of the cyst. The importance of this need to be emphasizes and a preoperative thorough discussion with the parents in this regard is of paramount importance.

**REFERENCES**