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CASE REPORT

Megacystis secondary to congenital urethral stricture

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Abstract Functional bladder capacity at a certain age can be accurately estimated and expressed as a function of age. We present a 13-year-old boy with abdominal distention who was presumed to have ascites. Abdominal ultrasound revealed that the bladder was severely distended, and cystoscopy showed a short stricture in the bulbar urethra followed by a large bladder without obvious borders. Under general anesthesia, bladder capacity was 9250 cc. Reduction cystoplasty was performed. At the 2-year follow-up, he voided spontaneously with 20 cc post-void residual urine.

Introduction

The bladder is a hollow muscular organ that serves as a reservoir for urine. When empty, the adult bladder lies behind the pubic symphysis and is largely a pelvic organ. In infants and children, it is situated higher. When it is full, it rises well above the symphysis and can readily be palpated or percussed. When over-distended, as in acute or chronic urinary retention, it may cause the lower abdomen to bulge visibly [1].

Case report

A 13-year-old boy presented with abdominal distention for ascites management. The patient had complained of anorexia and malaise for 5 years. Family history for ascites was negative. He complained of weak stream voiding. There was no history of perineal, pelvic or abdominal trauma.

Abdominal paracentesis had been done but there was no definitive diagnosis. He was given Ceftriaxone and Aldacton for ascites management.

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On physical examination, abdominal distention was noted to be dull in percussion. External genitalia were normal appearing and the testes were in scrotum. Serum creatinine was 1.1 mg/dl; urine analysis and all other laboratory tests were normal.

Abdominal ultrasound revealed kidneys of normal size and echogenicity without any dysplasia or hydronephrosis, and that the bladder was severely distended. On abdominopelvic CT scan, a huge cystic tumor without calcification was seen that occupied the whole abdominopelvic space.

We performed cystoscopy which showed a moderate stricture, 5 mm in length, in the bulbar urethra; an internal urethrotomy was done, entering a large space without obvious borders. A Foley catheter was inserted and, in order to estimate the size and characteristics of the space, retrograde cystography was performed (Fig. 1). Unfortunately, we did not have access to urodynamic study at that time for various reasons.

The diagnosis was a huge bladder secondary to long-lasting bladder outlet obstruction, so a reduction cystoplasty was planned. Under general anesthesia, before the incision was made, the bladder was filled in retrograde fashion with normal saline. Surprisingly, the capacity was 9250 cc. Abdominal wall musculature was normal appearing. The bladder was entered extraperitoneally and reduction cystoplasty was performed (Figs. 2 and 3). After removal of all catheters, the patient was discharged on the 9th postoperative day and he was prescribed to perform clean intermittent catheterization.

The pathology report on the resected specimen was normal bladder wall with intraepithelial and subepithelial polymorphonuclear leukocytes, predominant eosinophils, and dilated and congested vessels. Ratio of collagen to muscle fibers was normal.

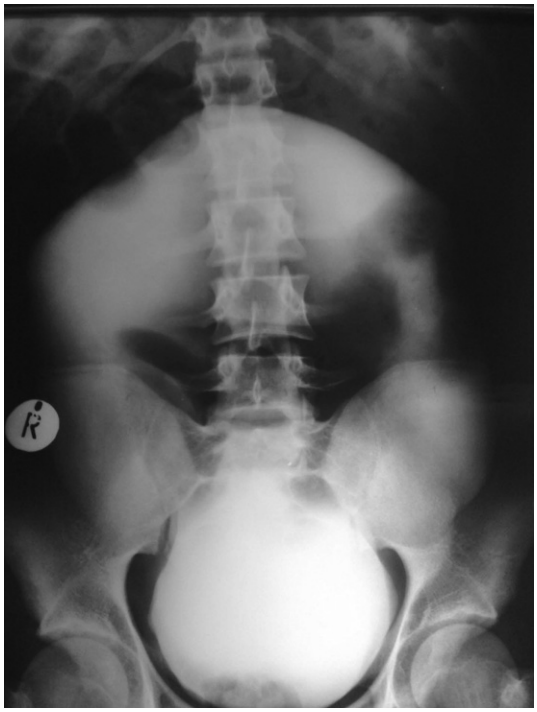

Figure 1 Retrograde cystography (note the upper border of contrast material).

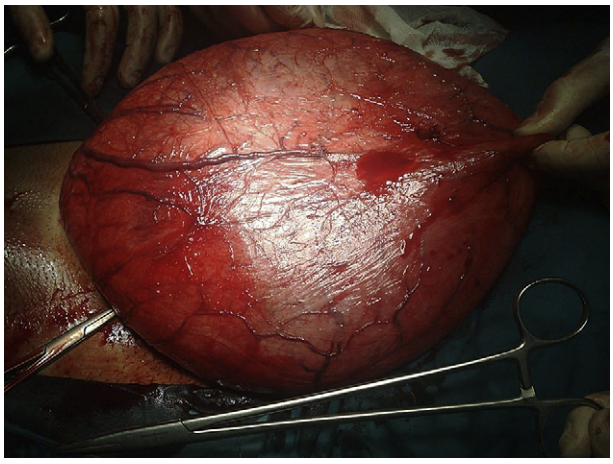

Figure 2 Intraoperative photograph of extraperitoneally released bladder.

After 2 years of follow-up the patient has no constitutional or urologic problems, voids spontaneously, and his post-void residual is 15—20 cc.

Discussion

The term megacystis is often used to describe any condition leading to a distended fetal bladder in utero without referring to the cause of the dilation. It has been recognized in association with particular conditions, as described below.

Prune belly syndrome represents a constellation of anomalies with variable degrees of severity. The three major findings are a deficiency of the abdominal musculature, bilateral intra-abdominal testes, and an anomalous urinary tract. The urinary tract is characterized by hydronephrosis, renal dysplasia, dilated tortuous ureters, an enlarged bladder, and a dilated prostatic urethra. Additional associated anomalies exist involving the respiratory tract, gastrointestinal tract, cardiac system, and musculoskeletal system. Various obstructive lesions of the distal

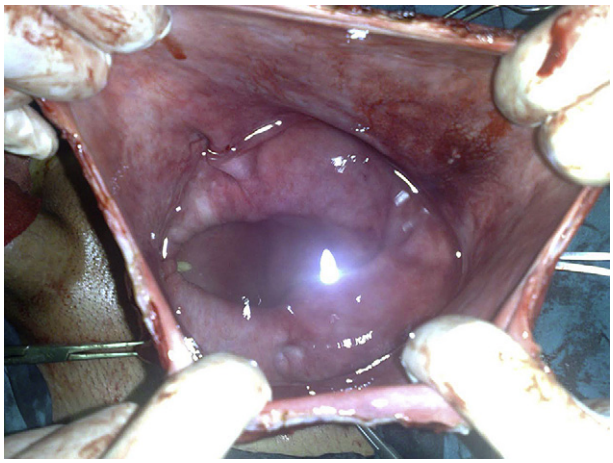

Figure 3 Intraoperative photograph of opened bladder (note tip of Foley catheter, bladder neck).

posterior urethra have been described, such as urethral atresia, valves, urethral stenosis, urethral membrane and urethral diverticulum, and are thought to occur in 20% of cases [2]. Incomplete forms of the syndrome would lack the typical abdominal wall features but have the common uropathy and cryptorchidism.

Berdon syndrome, also called megacystis—microcolon—intestinal hypoperistalsis syndrome, is characterized by megacystis with functional partial bladder obstruction, bilateral hydronephrosis, and associated microcolon with poor intestinal motility [3].

The association of a massively dilated bladder, hydroureteronephrosis, and bilateral vesicoureteral reflux has been termed the megacystis—megaureter association [4]. The constant recycling of bladder urine into the upper tracts effectively prevents true bladder emptying.

Patients with visceral myopathy and megacystis also have thin-walled bladders without trabeculation and diverticula suggestive of mechanical obstruction. Histologic analysis shows segmental intrafascicular fibrosis, elastosis and smooth muscle degeneration, but there is no inflammatory cell infiltration within the affected fascicles [5].

Regarding history and findings on physical examination, the absence of either urogenital anomalies, such as hydronephrosis, renal dysplasia, vesicoureteral reflux and undescended testis, or associated non-urological abnormalities and neurologic deficits, in addition to histopathologic findings and the patient's follow-up results indicating that he could void spontaneously after reduction cystoplasty, we think that we have managed a megacystis (perhaps the largest ever reported), and that the bladder decompensation occurred in response to a long-standing moderate urethral stricture. That being said, a rare variant of prune belly cannot be completely ruled out.

Conflict of interest

The authors declare that they have no conflict of interest.

References

[1] McAninch JW. Anatomy of the genitourinary tract. In: Tanagho EA, Mcaninch JW, editors. Smith's general urology. 17th ed. New York: McGraw-Hill; 2008. p. 6—7.
[2] Hoagland MH, Hutchins GM. Obstructive lesions of the lower urinary tract in the prune belly syndrome. Arch Pathol Lab Med 1987;111:154—6.
[3] Berdon WE, Baker DH, Blanc WA. Megacystis microcolon intestinal hypoperistalsis syndrome: a new cause of intestinal obstruction in the newborn. Report of radiologic findings in five newborn girls. AJR Am J Roentgenol 1976;126:957—64.
[4] Burbige KA, Lebowitz RL, Colodny AH. The megacystis-megaureter syndrome. J Urol; 1984:1131—3.
[5] Perk H, Serel TA, Anafarta K, Kosar A, Uluoglu O, Sarı A. Megacystis secondary to myenteric plexus pathology. Urol Int 2001; 67:313—5.