Isolated Congenital Megacystis with Spontaneous Resolution: An Exceedingly Rare Entity

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INTRODUCTION

Megacystis is a rare condition in infants, which is usually associated with refluxing megaureters, prune-belly syndrome (PBS), infravesical obstruction or presents as the megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS). Rarer even, is isolated congenital megacystis (ICM). The etiology of this entity is unknown. Myenteric plexus pathology[1] and a mild form of MMIHS[2] have been proposed as plausible pathologies. Here we add another case that resolved spontaneously.

CASE REPORT

A full-term 4-day-old neonate boy was referred to our center because of delayed and infrequent voiding. His fetal ultrasound had shown a distended, thin walled bladder without hydroureteronephrosis (HUN) and oligohydramnios at week 32. He passed meconium soon after birth, however did not void during the first 24 hours. After this period, voiding started but infrequently (three to four times a day). Ultrasound study of abdomen showed a huge bladder with 160 mL of volume. No HUN or increased bladder wall thickness was observed. Serum creatinine was in the normal range throughout the follow-up. Urine culture was negative and

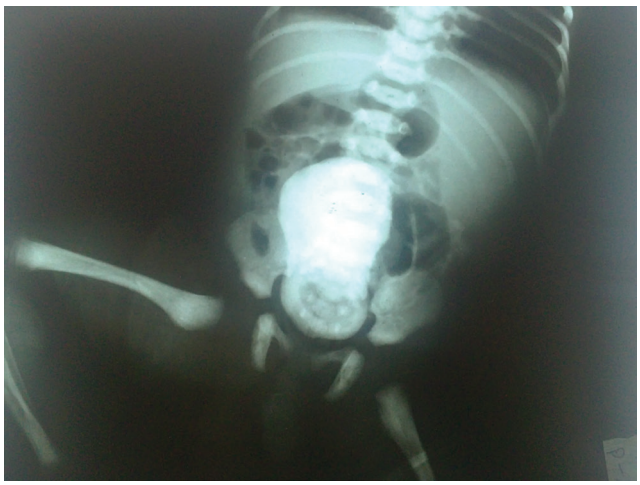

Figure .Voiding cystourethrography demonstrates a huge bladder without trabeculation or reflux.

the patient did not develop urinary tract infection (UTI) during the follow-up period. Physical examination showed no abnormality in the genital, rectal and sacral areas. Neurological examination was also normal.

Because of the lack of any signs of bowel obstruction, the pediatric gastroenterologist did not request barium enema or rectal biopsy. During performing the voiding cystourethrography (VCUG), the baby did not void up to the volume of 150 mL and the cystogram showed a huge bladder without trabeculation or reflux (Figure). The radiologist stopped the test at this volume because of concern regarding iatrogenic bladder rupture. Therefore, there was no urethrogram for the evaluation of the urethra.

Since the bladder wall was smooth and no HUN was present, we decided not to perform cystoscopy. Instead, we advised the parent to start clean intermittent catheterization (CIC). They were reluctant to accept this suggestion. Therefore, we advised them to come for frequent follow-up while on antibiotic prophylaxis. Serial ultrasound showed progressive shrinkage of the bladder, reducing to 80 mL at the age of six months. Bowel habit was still normal.

DISCUSSION

Reported cases of ICM are very few with different clinical course and management strategies.

Inamdar and colleagues reported a case of "vesical gigantism" that was initially managed by bladder catheterization, then cutaneous vesicostomy because of recurrent bladder distention, and later, by reduction cystoplasty due to high post-voiding residual urine.[3] They believed that this entity may "results from an expansion of the portion of the urinary bladder that develops from the allantois".

Shsimizu and colleagues reported another case that was managed expectantly up to the age of four years, however, the child developed UTI at that age. A hypocontractile detrusor and increased compliance were the urodynamic findings. Rectal biopsy revealed hypoganglionosis of the submucous and myenteric plexuses without thinning of the longitudinal muscle and connective tissue proliferation. They managed the patient by CIC and proposed that she might have a mild form of chronic intestinal pseudo obstruction syndrome.

Recently another case report appeared in the literature, presenting a case of ICM that resolved by the age of one year and remained asymptomatic afterwards without any therapeutic intervention.[4]

Since ICM is a very rare entity, when a baby is born with megacystis, other more prevalent pathologies should be ruled out first. PBS is diagnosed by the classic presentation of abdominal musculature defect and undescended testes. Infravesical obstruction could be diagnosed by a VCUG. A complete neurological and gastrointestinal evaluation is also mandatory to look for any signs of intestinal obstruction. Urodynamic study could demonstrate detrusor hypocontractility, if present. If there is no other anomaly, the diagnosis of ICM is suggested. Management of these patients should be individualized based on the presence of urinary retention, high post-voiding residual urine and UTI.

CIC, vesicostomy and observation with administration of antibiotic prophylaxis are all viable options. Regardless of the chosen therapeutic option, long-term follow-up is mandatory, because the clinical course of this entity is not fully understood.

CONCLUSION

The clinical course in this case highlight the fact that not all cases of ICM need extensive work-up and when the clinical setting justifies, cystoscopy, urodynamics and repeated VCUGs could be avoided.

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