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CASE REPORT

Spontaneous resolution of isolated congenital megacystis: The incredible shrinking bladder

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Abstract Isolated congenital megacystis represents a rare variant of fetal megacystis without other associated anomalies. The etiology is unclear, and various management strategies have been proposed. We report a case of isolated congenital megacystis that resolved over the first year of life with observation alone. Considerations for the evaluation and management of this rare entity are discussed.

Introduction

Isolated congenital megacystis (ICM) is an extremely rare condition of unclear etiology. Postulated pathologic mechanisms include a visceral myopathy [1] and a mild variant of megacystis-microcolon-intestinal hypoperistalsis syndrome (MMIHS) [2]. Due to the rarity of this condition, the optimal strategy for management is unclear, with previous authors reporting reduction cystoplasty [3] and clean intermittent catheterization (CIC) [2,4] as two options. Here, we report a case of ICM that resolved spontaneously over the first year of life.

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Case report

Prenatal course

A 29 year-old G3P2 healthy woman with no significant past medical or obstetric history was referred to our institution for evaluation of an enlarged fetal bladder with bilateral hydronephrosis. Prenatal ultrasound at 18 weeks gestation demonstrated a markedly enlarged fetal bladder ($2.0 \times 2.0 \times 2.5$ cm), moderate bilateral pelviectasis (4 mm AP diameter), the suggestion of a keyhole sign indicating a possibly dilated posterior urethra, normal amniotic fluid volume, and a normal-appearing three-vessel cord (Fig. 1). Due to the concern for a condition such as severe bladder outlet obstruction due to posterior urethral valves (PUV), our group recommended close ultrasonographic follow-up over the remainder of the pregnancy. Amniocentesis revealed normal XY karyotype, and the patient underwent

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serial ultrasounds for monitoring of the fetus. Over the course of the pregnancy, the fetal bladder remained large but thin-walled without diverticula and the testes were seen to positioned in the scrotal sac (Fig. 2), essentially ruling out prune belly syndrome (PBS) as an etiology for the enlarged fetal bladder.

Postnatal evaluation and management

The patient was born at 40 weeks via uncomplicated spontaneous vaginal delivery and weighed 4.5 kg. He was admitted to the neonatal intensive care unit and prophylactic antibiotics were started. He was initially allowed to void spontaneously, but ultimately a urethral catheter was placed due to elevated post void residuals. Creatinine after birth was 0.9 mg/dL, likely reflecting maternal levels. Postnatal ultrasound was notable only for an enlarged bladder; the hydronephrosis that was seen prenatally had completely resolved. A voiding cystourethrogram (VCUG) confirmed the finding of a markedly enlarged bladder, but showed no evidence of vesicoureteral reflux, and no evidence of PUV (Fig. 3).

After the VCUG, the patient was able to void and was discharged to home on prophylactic antibiotics. He was

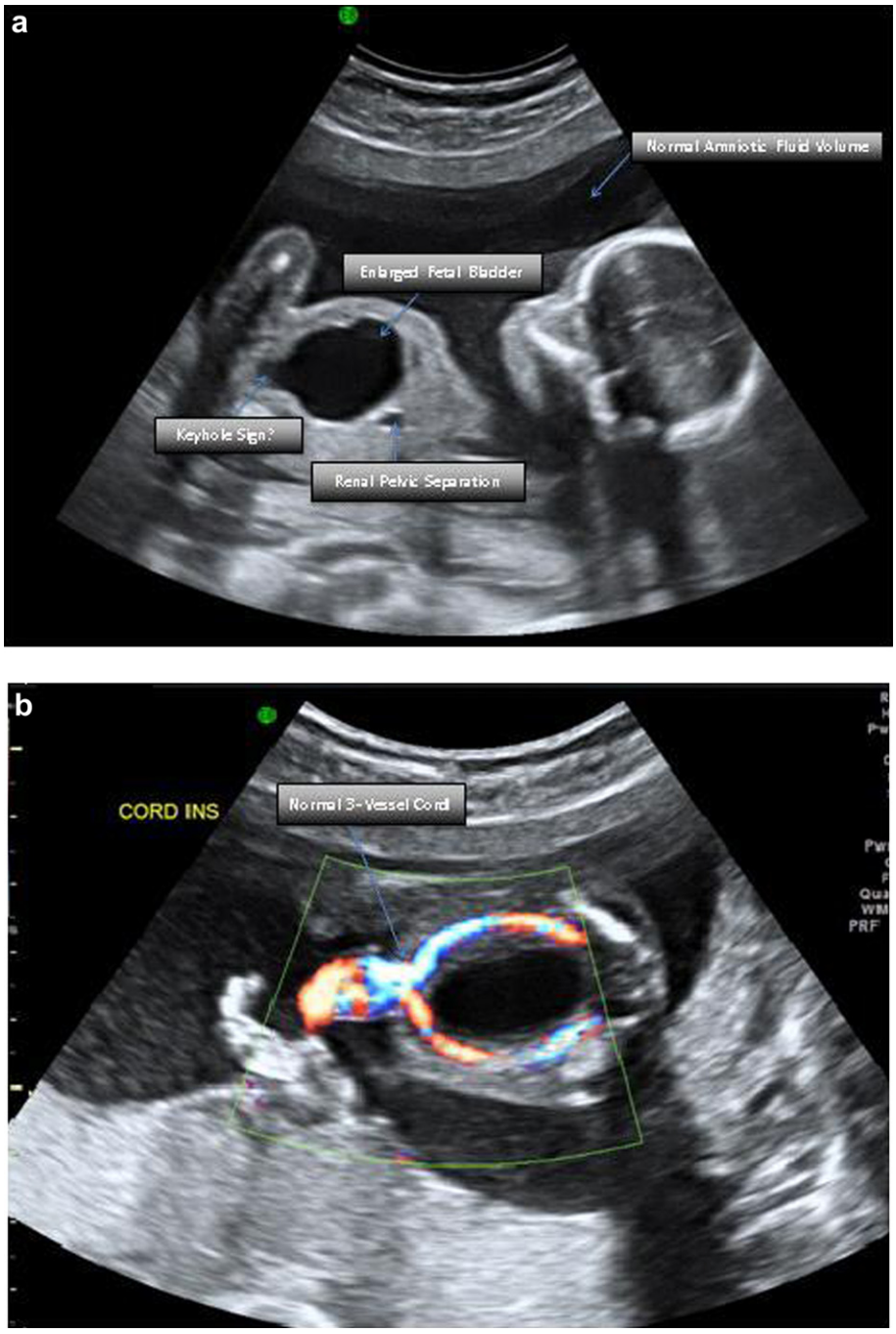

Figure 1 a. Prenatal ultrasound at 18 weeks gestation. b. Prenatal ultrasound at 18 weeks gestation with power doppler signal.