A Case of Prenatally Detected Bilateral Paraureteral Diverticula Associated with Neurofibromatosis

I. Iwao Yamagiwa
K. Kazuya Obata
H. Hiroyuki Saito
T. Takayuki Ouchi

Second Department of Surgery, Yamagata University School of Medicine, Yamagata, Japan

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Abstract
Although paraureteral diverticulum is thought to be a congenital anomaly, no prenatally detected cases have been reported. Herein, we report a case of bilateral paraureteral diverticula prenatally detected by maternal ultrasonography. This case also had dysplastic kidneys in association with neurofibromatosis.

Iwao Yamagiwa, 2-2-2 Iida-nishi, Yamagata 990-23 (Japan)

Paraureteral diverticulum is now well documented in children and thought to be a congenital failure of normal muscle development around the ureterovesical junction with subsequent herniation [1,2]. There have been many reports of cases with paraureteral diverticulum but few of them were diagnosed in the neonatal period [3,4]. Herein, we report a case with bilateral paraureteral diverticula that was detected in retrovesical cystic masses by maternal ultrasonography.

Case Report
A baby boy with an antenatally detected cystic mass in his pelvis was delivered at 38 weeks of gestation by cesarean section from a mother with neurofibromatosis. The cystic masses were detected at 33 weeks’ gestation. He also had many café-au-lait spots on his skin. A CT scan and an ultrasonograph revealed two retrovesical cystic masses changing in size with time. Small-sized kidneys were seen with hypo-enhancement of the contrast media in an enhanced CT. During a cystogram, the bladder was first filled with a contrast medium in normal appearance. Then both diverticula were abruptly filled with a contrast medium with massive vesicoureteral reflux. The sizes of the diverticula were as big as the bladder and both ureters were extremely tortuous (fig. 1). The serum creatinine level elevated to 2.7 mg/dl on his fifth day of age, then it gradually decreased and remained within a normal range. The baby was followed up at the outpatient clinic with the administration of antibiotics.

A cystoscopy performed at 5 months of age revealed a normal appearance of urethra and bladder including the trigone. The cystoscope was easily inserted into both diverticula. The mucosa of the diverticulum was similar to the bladder mucosa. A cystogram showed similar findings to the previous one and a slight vesicoureteral reflux was still observed (fig. 2).
Although the serum creatinine level still remained within the normal range, BUN levels gradually increased to 46 mg/dl at 10 months of age. Therefore, a decision was made to excise the diverticula.

In surgery, bilateral paraureteral diverticula were seen in the posterolateral portion of the bladder on each side. The wall of the diverticula was thinner than that of the bladder. The two diverticula were excised extravesically including about 1 cm of the ureter on both sides. The original ureteral orifices were sutured in the mucosal and muscular layers. Politano-Leadbetter type ureterocystoneostomies were performed on both sides. The postoperative course was uneventful and the renal function has remained normal for 2 years after the operation.

A histological examination revealed the diverticula to be lined with partly eroded transitional epithelium. The diverticular wall mainly consisted of fibrous tissue with scattered muscular bundles. The muscular bundles were partly hypertrophied. No inflammatory changes were observed.

Discussion

Three types of bladder diverticula in children are described: congenital, acquired and postsurgical [3]. Paraureteral diverticulum is considered to be a congenital muscular anomaly around the ureteral hiatus [1,2]. This is seen in infants, children, adolescents and adults, but is rare in neonates [3, 4]. Presenting symptoms include urinary tract infection, hematuria, urinary incontinence, vesicoureteral reflux, dysuria and occasionally obstruction [5]. This anomaly is usually solitary. This particular case was prenatally detected by maternal ultrasonography and had huge diverticula on both sides. We could not find a case with paraureteral diverticulum diagnosed prenatally in the literature. Bladder diverticula also occur as a part of some syndromes. They have been reported in patients with prune-belly syndrome, Ehlers-Danlos syndrome, kinky hair syndrome, Williams syndrome,
and cutis laxa [3, 5]. We could not find a case of paraureteral diverticulum combined with neurofibromatosis in the literature. This combination is thought to be incidental.

References


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