

Case Report

Unilateral hypoplastic kidney and ureter associated with diverse mesonephric remnant hyperplasia

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Abstract: Mesonephric remnants have been rarely reported in the genitourinary system and sometimes impose a diagnostic challenge both clinically and pathologically. We reported a case of mesonephric remnant hyperplasia with mixed acinar/tubular and epididymis/vas deferens-like morphologies occurring in the renal parenchyma of a unilateral hypoplastic kidney, which has not been previously described.

Keywords: Mesonephric remnant, kidney, hypoplasia

Introduction

Mesonephric ducts (also known as Wolffian ducts) are a pair of embryonic organ. In male, portions of each duct differentiate into epididymis, vas deferens, and seminal vesicles under the influence of testosterone. In female, the structures usually regress completely [1, 2]. Not infrequently mesonephric rests or remnants can be found in both male and female pelvic organs. In male, the mesonephric remnants persist as appendix of the epididymis and paradidymis in efferent ductules. In female, the mesonephric remnants can present as epoophoron and Skene's glands as well as Gartner's duct or cyst [1-3]. Rare locations of the mesonephric remnants have been reported in male, including renal pelvis, spermatic cord, vas deferens, urethra and prostate [4-7]. These inclusions/remnants usually represent innocent traces of the development process without association with structural abnormalities of the organ where they reside. Although it has been described in the renal pelvis in a case report, mesonephric remnant inside renal parenchyma has never been reported. Herein we described a case of renal medullary mesonephric remnant hyperplasia with mixed tubular and ductal morphologies and in association with unilateral renal and ureteral hypoplasia.

Clinical history

This was a 33 year-old male patient who presented with a new onset of gross hematuria 3 months ago. He was admitted to the emergency room. CAT scan of the abdomen and pelvis revealed a severely atrophic right kidney with remarkably dilated proximal hydroureter and mid ureter with thickened wall. It was felt by the radiologist that this was ureteritis and presence of a mass lesion in the right ureter could not be excluded. Cystoscopy retrograde pyelogram demonstrated that the right ureter (presumed to be hypoplastic) with severe stricture in the mid-distal portion and remarkable proximal hydroureter. The right kidney was very small and atrophic/hypoplastic with faintly visualized renal pelvo-calyceal system. The function of the right kidney was less than 2%. No imaging abnormalities were found in the left kidney and ureter. His hematuria stopped on the second day as he was pushing fluids. No imaging examination was recorded prior to this admission. He denied flank/abdominal pain, fever, and urinary difficulty. He had no history of frequent urinary tract infection, prior gross hematuria, urinary incontinence or stone disease. A right robotic nephrectomy was performed. The postoperative course was uneventful and he was discharged to home one day after surgery.

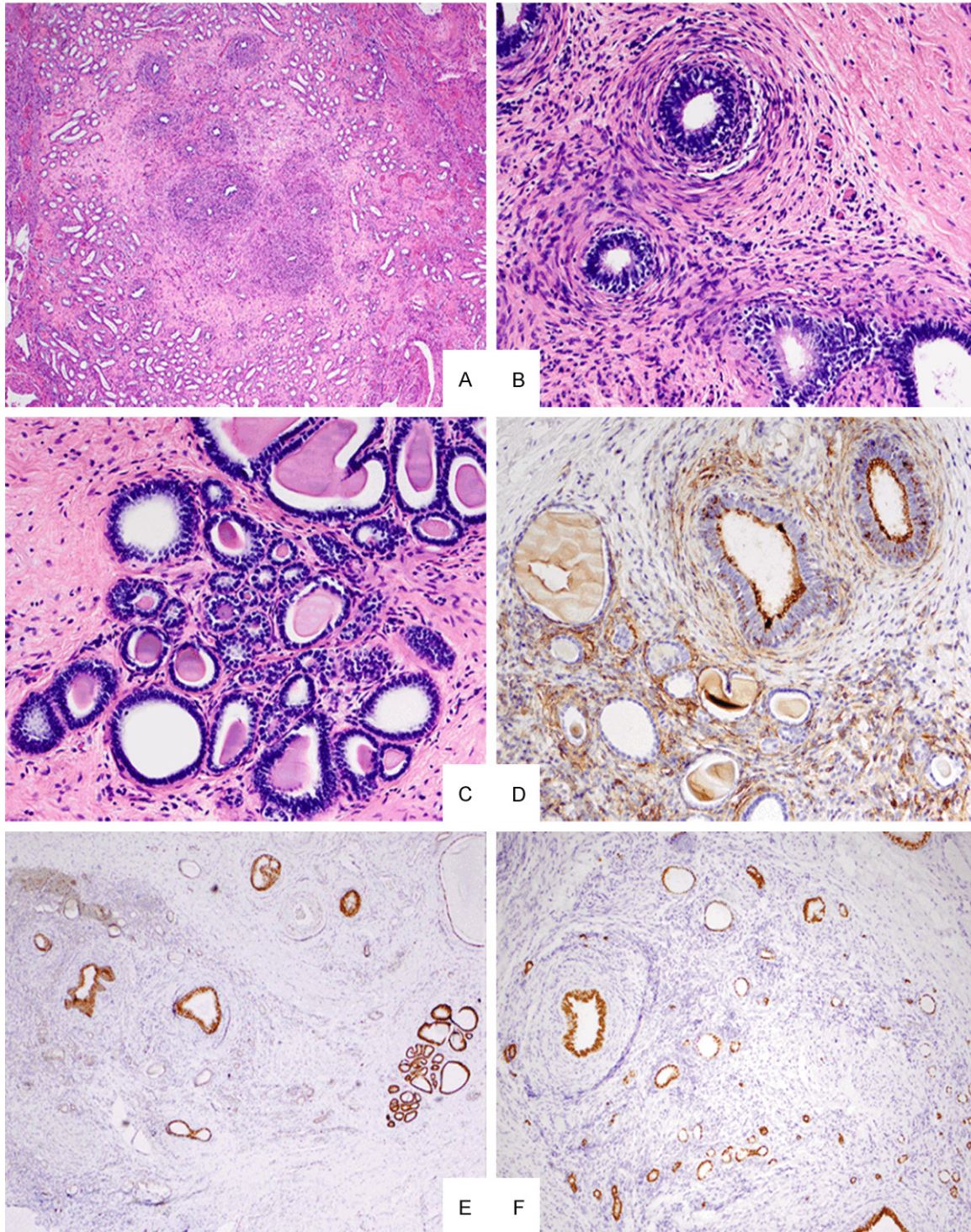


Figure 1. A. Mesonephric remnants are organized in a lobular pattern and located in renal medulla. B. Epididymis/vas deferens-like mesonephric remnants are surrounded by a coat of dense and cellular stroma. C. Acinar/tubular type mesonephric remnants are rounded in shape and variable in size with intraluminal colloid-like material. D. CD10 stains the brush border of epididymis/vas deferens-like ducts and surrounding stroma. Acinar/tubular type mesonephric remnants are negative for CD10. E. GATA3 immunoreacts with the epididymis/vas deferens-like epithelium and some of the acinar/tubular type epithelia. F. PAX8 is immunoreactive with both epididymis/vas deferens-like and acinar/tubular type epithelia.

Table 1. Immunohistochemical profiles of the mesonephric remnants

Mesonephric remnants	CD10	GATA3	P63	PAX8	ER	SMA	WT-1
Acinar/tubular type epithelium	-	Some + Some -	-	+	-	-	-
Epididymis/vas deferens-like epithelium	+ (brush border)	+	+ (basal cell only)	+	-	-	-
Remnant-associated stroma	+	-	-	-	+	+	-

+, Positive. -, Negative.

Material and methods

A right radical nephrectomy specimen with associated surrounding soft tissue was received. The specimen contained a 15 cm in length and 2.7 cm in diameter proximal dilated ureter. The distal ureter displayed a stricture, the lumen of which was less than 0.2 cm in diameter. The ureter had a tan-white, slightly granular, and glistening urothelium with focal areas of congestion. Sectioning of the specimen revealed a 6 × 1.5 × 1.5 cm atrophic/hypoplastic kidney. The renal parenchyma was predominantly gray-white and granular with normal appearance. The cortical-medullary junction was ill-defined and tan-brown to white. Representative sections were submitted for histologic examination.

Results

Microscopically the renal cortical parenchyma was remarkably thin and hypoplastic, with otherwise normal anatomic and histologic nephrons. The medulla was also markedly attenuated and distorted by multiple foci of aberrant ductal as well as aberrant tubular/acinar structures which were organized in a lobular fashion (**Figure 1A**). These aberrant elements occupied half of the medulla and extended into the renal pelvis. The epithelial linings of the ductal structures were composed of a layer of ciliated short columnar cells with amophilic cytoplasm. The nuclei were oval to elongated and uniform without atypia. Underneath was a layer of small basal cells with scant clear cytoplasm. This morphology resembled that of epididymis and vas deferens. Most of these epididymis/vas deferens-like ductal structures were surrounded by a coat of dense and cellular stroma (**Figure 1B**). There were also scattered aberrant acinar/tubular structures, which were lined by single layer of un-ciliated uniform small cuboid cells with pale to amophilic cytoplasm. These aberrant acini/tubules were frequently round in

shape but variable in size and arranged in a lobular growth pattern with intraluminal colloid-like material (**Figure 1C**). Some of them were also associated with dense and cellular stroma. The ureter was hypoplastic with focal muscular hypertrophy. No mass formation or heterogeneous mesenchymal elements was identified in the kidney and ureter.

On immunohistochemistry, CD10 was strongly reactive with the brush border of the epididymis/vas deferens-like epithelia as well as the stroma and non-reactive with the acinar/tubular type epithelia (**Figure 1D**). ER and SMA only reacted with their associated stroma. P63 was only reactive with the basal cells in the epididymis/vas deferens-like epithelia. GATA-3 reacted with the epididymis/vas deferens-like epithelia and some of the acinar/tubular-type epithelia (**Figure 1E**). Both the epithelial and stroma cells of these aberrant structures were negative for WT-1. PAX8 was strongly reactive with both the epididymis/vas deferens-like and acinar/tubular type epithelia (**Figure 1F**). These immunohistochemical results supported their mesonephric origin, and in conjunction with the histomorphology, a diagnosis was made as: hypoplastic kidney and ureter with medullary mesonephric remnant hyperplasia with mixed acinar/tubular and epididymis/vas deferens-like morphology. The immunohistochemical results are summarized in **Table 1**.

Discussion

Mesonephric ducts play an imperative role in the development of the urinary and male reproductive system, which include the ureter, renal pelvis, collecting tubules, ductus deferens, ejaculatory ducts, and seminal vesicle. Most of the embryonic mesonephric ducts are eventually replaced by metanephric ducts to form the permanent kidney. Although rarely, mesonephric remnant has been reported in the renal pelvis, spermatic cord, vas deferens, urethra,

prostate and prostatic urethra [4-7]. Most of the reported mesonephric remnants are composed of small acini/tubule lined by cuboidal to low columnar epithelium. More recently epididymis-like mesonephric remnants have also been reported in the prostate [8, 9]. In this case study, we described a mixed acinar/tubular and epididymis/vas deferens-like mesonephric remnant proliferation in association with a unilateral hypoplastic kidney and ureter, which has not been documented before. These two distinct mesonephric elements likely represented embryonic residues at different development stages.

Congenital anomalies of the kidney and urinary tract represent a broad range of disorders that result from abnormal embryonic development. These anomalies could be due to renal parenchymal malformations, abnormalities in renal migration, or abnormalities in the developing collecting system. The causes of congenital urinary anomalies can be either genetic or environment-related. Congenital anomalies of urinary system usually occur in children, but if unilaterally, it can be unnoticed until adult age when symptom develops. The frequency of unilateral small kidney has been reported to be one in 500 autopsies [10]. A kidney may be small because of congenital hypoplasia or pyelonephritic shrinkage or as a combination of both. Simple renal hypoplasia, which consists of a lower number of structurally normal nephrons, is distinct from renal dysplasia, which displays renal parenchymal malformations. In hypoplastic kidneys, in spite of small in size and number, the components of the calyceal system present a normal functioning and keep a relationship with the volume of the parenchyma [10], although it remains unknown what causes the renal hypoplasia, genetic determinants are thought to play a role. Other factors, such as vitamin A deficiency, have also been associated with urogenital malformations and renal hypoplasia [11]. Hypertension and urinary tract infection have been reported to be common in renal hypoplasia, but these were not present in our case.

Although in this case the relation between mesonephric remnant hyperplasia and hypoplastic kidney and ureter is unclear, since the renal cortex and the medullary collecting system have different embryonic origin [12], they

might be just an incidental concurrent process, however, it cannot be excluded that the development of hypoplasia might have affected the regression process of some portion of the mesonephric ducts. Renal hypoplasia must be differentiated from acquired atrophic kidney, which is small and contracted. In this case, since parenchyma atrophy is not present, mesonephric remnant as a cause of the small kidney seems unlikely, but it certainly could have impaired the renal function.

Although rare, intrarenal parenchyma mesonephric remnant should be differentiated from neoplastic process, including metanephric stromal tumor, metanephric adenofibroma congenital mesoblastic nephroma, and Wilms tumor. These neoplastic processes can usually be diagnosed by the presence or absence of a mass forming lesion and their typical histomorphologic features, including the presence of distinct epithelial as well as mesenchymal elements. Immunohistochemistry will also offer an additional aid in reaching the correct diagnosis.

Disclosure of conflict of interest

None.

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