First adult case of sporadic localized glomerulocystic kidney mimicking a tumor

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Received March 15, 2014; Accepted December 3, 2014

DOI: 10.3892/ol.2015.3060

Abstract. Glomerulocystic kidneys (GCKs) are mainly observed in infants and young children, and are characterized by the cystic dilatation of Bowman’s space to form glomerular cysts (GCs). GCKs are associated with various conditions. Additionally, the cystogenesis of GCKs remains controversial. The present study describes a rare adult case of a sporadic localized GCK that radiologically mimicked a multilocular cystic tumor, and analyses the features of GC. A 42-year-old male with hematuria underwent a right partial nephrectomy for a cystic mass. The majority of the cyst was distributed in the cortex and contained a single collapsed glomerulus. Using serial sections, narrow and serpiginous proximal tubules that continued to the GCs were detected. These findings suggested that obliteration at the glomerulotubular junction was not the primary cause of GC in this case. To the best of our knowledge, this is the first adult case of a sporadic localized GCK mimicking a tumor. Unnecessary surgical procedures may be avoided by careful evaluation of computed tomography scans and magnetic resonance imaging, although localized GCKs are quite rare.

Introduction

Glomerulocystic kidneys (GCKs) are characterized by the cystic dilatation of Bowman’s space to form glomerular cysts (GCs), and are mainly observed in infants and young children in association with the following conditions: Hereditary polycystic kidney disease, tuberous sclerosis, renal dysplasia and renal ischemia, and certain medications, including lithium (1). To the best of our knowledge, only 35 cases of GCKs have been reported in adults worldwide (1-3). The majority of these patients presented with decreased renal function and subsequently received hemodialysis treatment, however they eventually progressed to end-stage renal disease (2,3). However, a few cases had normal renal function or were asymptomatic (1-4). GCKs are generally diagnosed by open renal biopsy. The lesion is not generally recognized as a neoplastic mass and cases of GCKs mimicking multilocular renal carcinoma are rare. The present study describes the first adult case of a sporadic localized GCK that presented as a cystic mass mimicking a neoplasm, and provides an analysis of the features of GC. Informed consent was obtained from the patient’s family.

Case report

An asymptomatic 42-year-old male presented to Saiseikai Senri Hospital (Osaka, Japan) was revealed to have microscopic hematuria following a medical check-up. The patient was revealed to have a localized nest of multilocular cysts, without any expansile nodules, which measured 2x4 cm in diameter. The cystic mass was detected in the lower pole of the right kidney by abdominal ultrasonography, contrast-enhanced computed tomography (CT) (Fig. 1A) and gadolinium-enhanced magnetic resonance imaging (MRI) (Fig. 1B). Contrast-enhanced CT revealed enhancement in the lesion [Bosniak classification category 3 (4)]. Cystic renal cell carcinoma could not be ruled out, therefore, a right partial nephrectomy was performed.

Grossly, the lesion was composed of multiple cysts filled with serous fluid, each measuring ≤8 mm in maximum diameter, and was distributed in the cortex of the resected kidney (Fig. 2). A small calculus measuring 2 mm in diameter was deposited in the outer medulla directly under the lesion. Microscopically, the majority of the cysts were lined by a single layer of flattened epithelium and a collapsed glomerulus was evident (Fig. 2C). In the corticomedullary junction, a few cysts lined by epithelial membrane antigen- and CK34βE12-positive cuboidal epithelium were suggestive of a derivation from the distal tubule or collecting duct. This
lesion was diagnosed as a sporadic case, as no family history or clinical history was found that was associated with GCKs.

A serial section study was performed for light microscopy to examine the GTJ. A total of 250 4-µm thick sections and 100 2-µm thick sections were stained with hematoxylin and eosin (HE) and Periodic acid-Schiff (PAS), respectively. A GC was defined as Bowman's capsule dilation of more than twice the diameter of a normal Bowman's capsule (320 µm), as described in a previous study (1). Each single specimen contained 50-80 cysts. Eight and six GTJs from 30 examined GCs were detected in these HE- and PAS-stained sections, respectively. These sections revealed a connection between the GCs and the proximal tubule, although the connected tubule became narrow and serpiginous (Fig. 3). The patient was lost to follow-up.

Discussion

Generally, GCKs diffusely involve the bilateral kidneys in infants and young children. To the best of our knowledge, this is the first adult case of a sporadic GCK mimicking a tumor. Retrospectively, the enhancement of this lesion that was identified on CT was determined to be that of a residual normal cortex. Radiologically, the lesion was almost entirely localized in the cortex, without renal surface deformity or protrusion from the kidney, and did not have the capsular and peritumoral change associated with invasive or expansive growth. These findings suggest that it was a non-neoplastic lesion. Unnecessary surgery may be avoided in future cases by careful evaluation of the CT and MRI, although localized GCKs are quite rare.
Obliteration at the glomerulotubular junction (GTJ) has been assumed to be the cause of GCKs associated with several diseases, however, the exact cause remains controversial (3,5,6). Hotta et al (5) used serial sections to demonstrate GTJ stenosis and suggested that periglomerular fibrosis induced stenosis of GTJ. By contrast, Liu et al (3) identified no stenosis or obstruction of the GTJ using serial sections. In the present case, a connection between the GCs and the proximal tubule was identified. These findings suggested that obliteration at the GTJ was not the primary cause of the GCs in the present case. Since this case was composed of large GCs with non-detectable surrounding proximal tubules, it was reasonable to assume that the GCs were composed of Bowman's capsule and part of the proximal tubule (7). We hypothesize that proliferation of the parietal cells and renal tubular cells, fluid accumulation and remodeling of the nephron (6) were the main causes of cyst development in the present case. As various conditions are associated with the formation of GC, such as hereditary polycystic kidney disease, tuberous sclerosis and renal dysplasia, we hypothesize that different pathogenic mechanisms of GC formation exist.

In summary, the present study reported an adult case of a sporadic localized GCK that resembled a cystic renal neoplasm, and provided analysis of its characteristic histopathological features using serial sections.

Acknowledgements

The authors would like to thank Mr. Masaru Nishino, Mrs. Noriko Yokozeki and Mr. Manabu Kobayashi (Department of Central Clinical Laboratory, Saiseikai Senri Hospital, Suita, Osaka, Japan) for their technical assistance in the serial sectioning.

References