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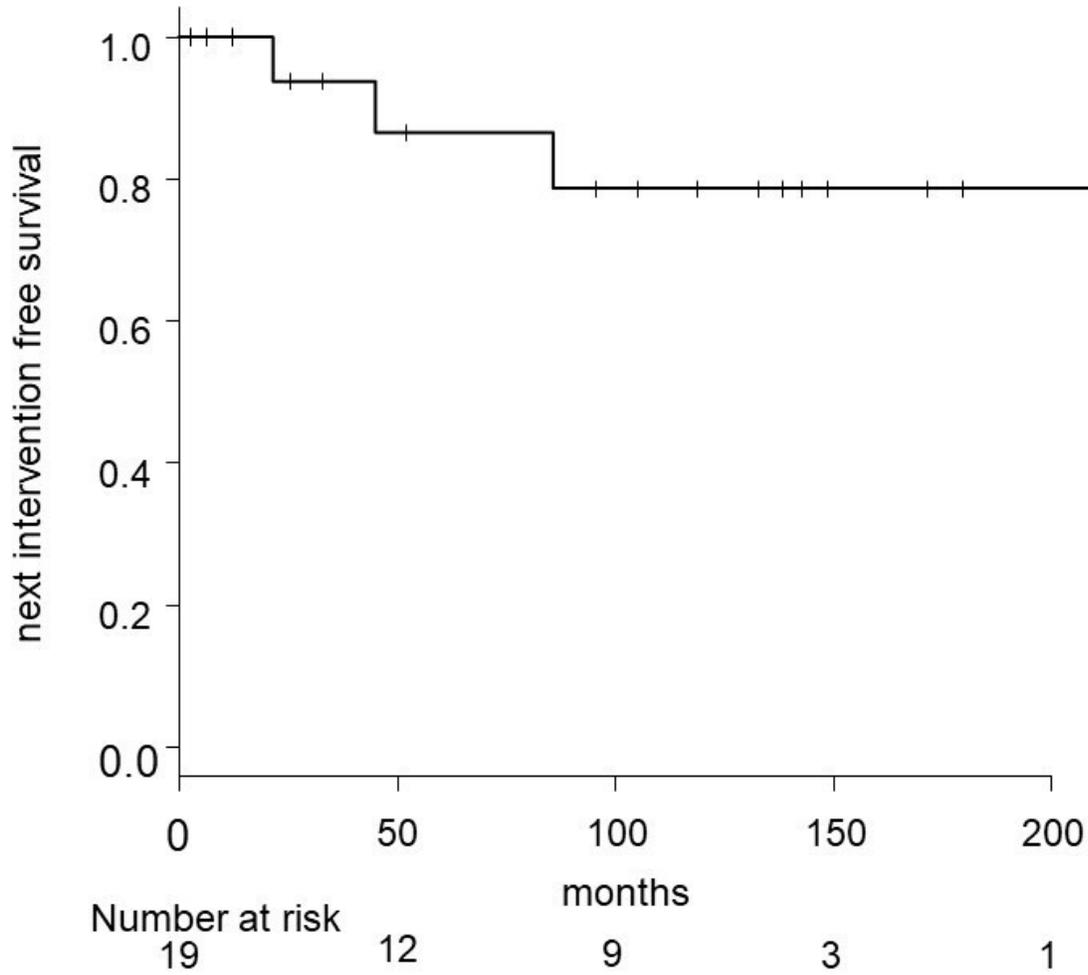
Introduction & Objectives: Renal cell carcinoma (RCC) in von Hippel-Lindau disease (VHL) patient is characterized by the high risk of multiple and bilateral occurrences throughout the life. Surgical interventions require both oncological and renal functional outcomes. We retrospectively evaluated the surgical management and outcomes of VHL associated RCC.

Materials & Methods: From 1993 to 2018, a total 17 VHL patients underwent renal tumor surgery. Clinicopathologic characteristics, surgical management, complications, and prognosis were retrospectively analyzed. Median follow-up was 102 months (range: 3-248 mo).

Results: Median age at surgery was 33.6 years. 17 patients underwent 21 kidney operations, including 17 open enucleations, 2 open partial nephrectomies, 1 laparoscopic partial nephrectomy, (1 to 9 tumors removed for a single kidney, median tumor size, 20 mm; range, 6-55 mm), and 1 radical nephrectomy. We basically intended to perform nephron sparing surgery (NSS) when the maximum tumor reached to 2.0 cm in size (2 cm rule). We also resected all suspected lesions, including small nodules and cystic lesions as possible as we can using intraoperative Dopplar sonography. Among 83 clear cell RCCs resected, 82 (99%) were histologically diagnosed as nuclear grade 1 and grade 2 and one (1.2%) with grade 3. Twenty two out of 44 lesions (50%) clinically considered as simple cysts according to preoperative imaging studies were histologically confirmed as cystic clear cell RCC (G1 and G2). Major surgical complications, including acute renal failure and blood transfusion, were not occurred. Local recurrences occurred in 4 renal units for 3 patients. The next intervention-free survival for 20 renal units which had underwent NSS was not reached and the estimate second intervention-free survival probabilities at 3 years, 5

years and 10 years were 93.8%, 86.5%, 78.7%, respectively (Fig.1). Renal cancer death was not observed.

Fig.1 The Kaplan-Meier curves analyzing the renal tumor recurrence greater than 2cm



Conclusions: The treatment strategy at 2 cm rule for VHL associated RCC seems to be acceptable both in oncological and renal functional outcomes and safety of surgical procedure. Resecting small tumors and premalignant lesions including renal cysts may contribute to better results. Early detection of the renal tumor and the optimal therapeutic interventions along with preserving renal function are thought to be important for patient with VHL disease.