American Urological Association
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AUA
VIRTUAL
EXPERIENCE

Clinical, surgical, pathological and follow-up features of kidney cancer in patients with Von Hippel-Lindau syndrome: novel insights from a large European consortium

Results

DEMOGRAPHIC FEATURES	
Patients	78
Gender	
Male	51.3%
Female	48.7%
Mean age at diagnosis	34.2
Mean age at renal management	41.1

RENAL FUNCTION	
Chronic kidney disease	
Preoperative	16.5%
Postoperative	25.3%

CLINICAL FEATURES	
Manifestations at VHL diagnosis	
Only renal	2.6%
Central Nervous System	61.5%
Pancreas	62.8%
Eyes	29.5%
Adrenal gland	14.1%
Others	16.7%
Central Nervous System + pancreas	42%

FINAL PATHOLOGY	
Mean tumor size (mm)	42
Tumor	
Small (<40mm) low-grade (G1-2)	52.4%
Large (>40mm) low-grade (G1-2)	22.6%
Small (<40mm) high-grade (G3-4)	11.9%
Large (>40mm) high-grade (G3-4)	13.1%

ONCOLOGIC OUTCOMES	
Mean follow-up (mo)	123
Median follow-up (mo)	83
Clinical progression at	
5 years	15%
10 years	36.6%
All-cause mortality	
5 years	5.2%
10 years	7.6%
Cancer-specific mortality	
5 years	3.4%
10 years	5.8%

Conclusions

- In Von Hippel-Lindau syndrome, virtually all patients with renal tumors have manifestations in other organs;

- Central nervous system (61.5%) and pancreas (62.8%) represent the most frequent VHL localizations;

- Despite more then 50% of renal VHL localizations are small and with low grade at final pathology, cancer-specific mortality is not neglegible.