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

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INTRODUCTION

Von Hippel-Lindau (VHL) syndrome is a relatively rare hereditary condition leading to renal and extrarenal manifestations. Although physiopathology and involved molecular pathways have been extensively analyzed, few data are available regarding the natural history, clinical and surgical aspects, complications and follow-up after renal surgery in this specific cohort of kidney cancer patients.

MATERIALS AND METHODS

A multi-institutional European consortium was built including Caucasian patients with VHL syndrome and kidney cancer treated at 8 referral institutions. Descriptive and survival analyses were performed.

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 the context of VHL patients, virtually all patients with renal tumors already have manifestations in other organs at first presentation. In candidates for renal treatment, CNS and pancreas are the most frequent extra-renal VHL localizations. Although the majority of tumors are small and of a low grade, cancer-specific mortality is not negligible.