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Von Recklinghausen's Disease and Pheochromocytoma: A Ten-Year Experience

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Background: Neurofibromatosis Type-1 (NF1) carries an increased lifetime risk of developing pheochromocytoma approximating 1 to 5%. Most experts recommend NF1 patients be screened for pheochromocytoma if they develop hypertension. Because of its rarity, the literature on NF1-associated pheochromocytoma consists primarily of single patient case reports.

Methods: Retrospective analysis of a prospectively collected database of all patients undergoing pheochromocytoma resection by a single surgeon from 2003-2012. NF1 and non-NF1 patients were compared. Statistical significance was tested with Fisher's exact test for categorical variables and the Wilcoxon rank sum test for continuous variables.

Results: Of 56 patients undergoing resection of pheochromocytoma, 6 (11%) had clinical and familial features of NF1. Overall, the mean age at presentation was 51 years (range 19-75) and 38% were males with no statistically significant difference between NF1 and non-NF1 patients. All 6 (100%) NF1 patients had pheochromocytoma diagnosed incidentally during work-up for another condition, while 28/50 (56%) non-NF1 associated pheochromocytomas were diagnosed incidentally ($p=0.071$). Hypertension was present in 1 (17%) NF1 patient, whereas 37 (74%) of the non-NF1 patients had hypertension ($p=.011$). Tumor size was significantly smaller in NF1 compared to non-NF1 patients (mean tumor dimension 2.78 cm vs. 5.66 cm, respectively, $p=0.014$). The two groups did not differ significantly with respect to race, ethnicity, symptoms, use of endoscopic adrenalectomy, mean procedure time, or occurrence of complications.

Conclusions: Although NF1 patients have a well-known increased risk of developing pheochromocytoma, in the current series all NF1 patients referred to the surgeon for adrenalectomy had pheochromocytoma diagnosed incidentally. Nevertheless, NF1 patients had significantly smaller tumors than other patients treated for pheochromocytoma, perhaps due to higher frequency of imaging occasioned by their other neoplasms. The common recommendation to screen for pheochromocytoma when hypertension develops would have failed to spur screening in 83% of these NF1 patients.

Variable	NF group (n=6)	Non-NF group (n=50)	p
Mean Age in years	50.8	50.6	.926
Male	3 (50%)	18 (36%)	.661
Race White Black Pacific Islander	 6 0 0	 40 9 1	.620
Hispanic Ethnicity	1	8	>.999
Symptoms	3	23	>.999
HTN	1	37	.011
Incidental	6	28	.071
Leaves OR on Pressors	3	15	.374
Surgical Procedure laparoscopic retroperitoneoscopic open converted to open	 4 1 1 0	 18 12 19 1	.538
Minimally Invasive	5	30	.393
Mean EBL (mL)	100	312	.704
Mean RBC Transfused	0	.6	.223
Mean Tumor Size in cm	2.78	5.63	.014
Mean Procedure Time in min	142	178	.617
Complications	0	9	.575