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EDITORIAL COMMENT

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These authors report 1 of the largest studies in the world aimed at identifying factors predictive of malignant PCC. Unlike most other solid tumors, malignant PCC is only defined by metastasis at sites where chromaffin cells are normally absent. Thus, much effort has been devoted to identifying reliable histological and molecular markers to predict the behavior of resected PCC and tailor appropriate followup. While the adrenal gland PCC score was advocated by Thompson¹ and others, some groups noted poor discriminatory power to distinguish benign from malignant lesions.^{2,3} Clearly these authors provide a few clinical parameters and specific features that may improve the identification of malignant PCC. Combining these variables and generating an ROC can allow quantitative comparisons to the adrenal gland PCC scale score.¹

Notably the study excluded patients with a known history of a hereditary PCC syndrome. Nevertheless, even in those without a familial history the importance of genetic causes cannot be overemphasized. In a study by Neumann et al 66 of 271 patients (24%) who presented with nonsyndromic, nonfamilial PCC had genetic mutations that were potentially responsible for PCC development.⁴ More importantly mutations of the SDH gene, subunit B are associated with greater than 70% of malignant PCCs.⁵ Thus, at our institution we frequently perform SDH germline testing when patients present with PCC at an early age or there are any aggressive histological features. In the future it is likely that combining genetic/molecular testing and clinicopathological characteristics will allow us to identify patients with malignant PCC for aggressive surveillance and/or enrollment in adjuvant clinical trials.

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