

Preface

Adrenocortical carcinoma (ACC) is a disease that most physicians, including many endocrinologists, will rarely, if ever, diagnose or let alone treat during the course of their medical practice. Medical textbooks of endocrinology and oncology rarely dedicate an entire chapter to this disease entity. The pursuit of research and clinical excellence in uncommon diseases is extremely challenging because of a lack of research prioritization, nonexistent treatment guidelines and overall paucity of coordination between researchers and physicians. ACC is one such disease where no infrastructure for a unified research agenda and no consensus treatment guidelines had been developed.

While a number of international meetings over the past two decades have indeed focused on adrenal tumors in the context of hormone excess (Cushing's syndrome, hyperaldosteronism, and pheochromocytoma), few have exclusively catered to the science and clinical care of ACC and those afflicted with the disease. With only a single FDA-approved drug for ACC (mitotane – a derivative of the pesticide DDT), institutional experiences varied widely until recently when historic biases have slowly yielded to data-driven treatment strategies. A large part of the impetus for this push has come from Europe, where the availability of country-wide integrated networks for treatment has allowed a small number of centers in Italy, France, and Germany (among others) to develop specific expertise and specific treatment protocols for this rare disease.

In an attempt to facilitate coordination of global efforts, a consensus conference was organized and held at the University of Michigan in September 2003. At that meeting, an international group of physicians and scientists with research interest and clinical expertise in ACC set up initial guidelines for the diagnosis and treatment of ACC. Three principles emerged. Successful treatment of ACC demands coordinated care in the context of a multidisciplinary team dedicated to the disease. Future therapies for ACC need to be predicated on hypothesis-driven research based on a thorough analysis of tumor biology. Lastly, major advancement in the field demands national and international collaborative networks to facilitate analysis of large datasets and coordinate future clinical trials. The FIRM-ACT (First International Randomized Trial in locally advanced and Metastatic Adrenocortical Cancer Treatment) that coordinated over 35 ACC centers in a single multinational trial set precedence for the actualization of these principles. The Second

International Adrenal Cancer Symposium: Clinical and Basic Science held at the University of Michigan in March 2008 built upon the momentum of the 2003 consensus meeting and the successful development of a large international ACC network through the FIRM-ACT trial.

Over the past decade, the ACC research community has grown to a critical mass with new data emerging in the laboratory and clinic. In times of electronic publications we routinely rely on journal articles and expert reviews on both clinical and research topics. While such publications are informative, when approached by Springer about the possibility of editing such a textbook, we became convinced that the time had come to compile the accumulated clinical and basic science knowledge of 50 years of active research on this rare cancer into a concise medical textbook. The overall goal of this book is therefore to provide definitive reference material for scientists and clinicians, to introduce trainees to concepts of ACC management, and to stimulate further research, future collaborations, and networking.

As opposed to a solitary review article, a textbook with multiple chapters dedicated to discrete topics in the field provides contributors the opportunity to objectively review historic data and detail the current state of clinical care and research accomplishments. While this is a major advantage of a textbook, it is also a major challenge for a book that focuses solely on a rare cancer where data are scant. In editing this book, we tried to ensure that each individual chapter covers well-established knowledge in the area, but also allows room for expert opinion. Lastly, because ACC has been linked to several genetic disorders that usually escape discussion in a focused review of adrenal tumors, the various syndromes will be discussed in their entirety in separate chapters. The 32 Chapters of the 9 Sections are authored by the scientific and clinical leaders in the field.

With publication of this first edition, the editors want to extend special thanks to our colleagues within the ACC community, the contributors, Rachel, Todd and Lesley of the editorial staff at Springer Publishing House, and Lisa K. Byrd of the University of Michigan.

We are hopeful that this first edition of the textbook provides an intellectual platform for the continued coalescence and dissemination of knowledge on ACC in future editions. Both the authors and editors welcome comments and recommendations for improvement in writing or via electronic mail. The editors' and authors' institutional and e-mail addresses are given in the contributor's section.

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