

Preface

Neuroendocrine tumors arise from cells dispersed throughout the body. Historically, they have been thought to be a group of very rare and indolent diseases capable of causing a variety of esoteric hormonal syndromes. Over the past decade, a number of major advances were made in our understanding of the epidemiology and molecular biology of these not-so-rare tumors. Although several studies have demonstrated a significant heterogeneity among neuroendocrine tumors by primary site and proliferative rate, recent analyses of the population-based registries confirm a consistent and continuing rise in its incidence. Further, because of the relative longer survival enjoyed by patients with this disease, it is now recognized that the prevalence of neuroendocrine tumors exceeds 100,000 individuals in the United States alone.

Neuroendocrine tumors are often well differentiated and associated with an indolent clinical course, but they can also present in much more aggressive forms. Due to their ability to produce hormones, their clinical presentations can be rather unusual and dramatic, requiring prompt expert treatment. Some neuroendocrine tumors are associated with genetic syndromes, which should be suspected particularly when the tumors arise at an early age or in family clusters.

While early stage neuroendocrine tumors can be cured by surgery, the disease is generally incurable when presenting with metastases. Despite the reputation of being indolent, most patients with advanced disease will eventually succumb to the disease. Successful management requires an understanding of the disease process as a whole and a multi-modality approach. Depending on the case, the inputs from medical oncology, surgery, endocrinology, gastroenterology, pathology, radiology, genetics, and nuclear medicine are required.

While our understanding of the molecular pathogenesis of neuroendocrine tumors remains incomplete, progress has been made. Studies of the MEN1 gene function have led to our understanding of its role in epigenetic regulation and control of endocrine cell proliferation. More recent studies have also demonstrated the importance of angiogenesis and the activation of the mammalian target of rapamycin (m-TOR) pathway in the genesis and progression of neuroendocrine tumors.

These advances have generated a renewed interest in the development of novel therapeutic options for neuroendocrine tumors using the novel molecular targeted agents. During the last few years, three of those targeted agents have been evaluated in pivotal randomized phase III studies for neuroendocrine tumors. Octreotide, sunitinib, and everolimus have successfully demonstrated significant antitumor activity against neuroendocrine tumors. These studies not only demonstrated that rigorous evaluation of antitumor agents in what was thought to be a rare disease is feasible, but also demonstrated that the integration of molecular targeted agents can lead to critical advances in the management of those patients.

In this volume, we have gathered an impressive array of thought leaders in the field from around the world. They have generously undertaken a comprehensive review of the epidemiology, biology, and management of neuroendocrine tumors. Recent advances in our understanding of molecular biology and emerging therapeutic options are emphasized.

We, as all the participating authors, hope that this work will help demystify some important misconceptions regarding neuroendocrine tumors, and that it may help to improve the treatment of patients and families affected by these diseases.

Good reading!

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