PREFACE

Paragangliomas and pheochromocytomas are highly interesting tumors that still represent a diagnostic and therapeutic challenge. Their organization is to a remarkable extent akin to that of the normal tissues of origin, that is, the paraganglia and the adrenal medulla. In fact, in some cases, their pathological effects are mediated by the uncontrolled release of physiological products of the paraganglia system, that is, catecholamines. Thus, paragangliomas and pheochromocytomas may present clinically with hypertension, tachycardia, hyperglycemia, and gastrointestinal effects. Prevention of the potentially life-threatening complications of excessive catecholamine release is therefore of primary relevance. An additional challenge of paragangliomas, and of some pheochromocytomas, is their unpredictable behavior. Mostly, these are slow-growing tumors that remain confined to the site of origin; however, they tend to relentlessly infiltrate into the adjacent tissues, a process that may complicate surgical resection, particularly for those paragangliomas that infiltrate the anatomically complex region of the skull base. Furthermore, for poorly understood reasons, an unpredictable minority of paragangliomas and pheochromocytomas exhibits metastatic capacity. The problem with these metastatic tumors, and with those that cannot be radically removed with surgery, is that systemic therapies are largely ineffective, while the efficacy of radiotherapy is still debated.

In the last two decades, exciting progress has been made in the field of paraganglioma and pheochromocytoma genetics. Several hereditary paraganglioma syndromes have been defined, and, although no new therapies have been developed based on these new data, it is now possible to identify, with some degree of reliability, subsets of patients that are at risk of more aggressive tumor behavior. In any case, treatment of paraganglioma and pheochromocytoma requires specialized knowledge and solid experience, which, given the rarity of these diseases, is available only in highly specialized centers.

This book includes specific chapters written by experts from several countries that deal with relevant aspects of paraganglioma and pheochromocytoma diagnosis and therapy. Chapter 1 addresses the genetic findings that are most relevant to diagnosis and management, chapter 2 deals with the biochemical diagnosis, chapter 3 with imaging, chapter 4 with the surgical and pharmacological management, mainly of trunk paraganglioma and pheochromocytoma, chapters 5 and 6 with the histopathological, genetic, and clinical characteristics of carotid body, and vagal and tympano-jugular paragangliomas.

We hope that these chapters will provide the reader an useful insight into important aspects of state-of-the-art diagnosis, management and treatment of paragangliomas and pheochromocytomas.

Renato Mariani-Costantini, MD
Professor of General Pathology
Department of Medicine, Dentistry and Biotechnology
"G. d'Annunzio" University, Chieti, Italy

Head, Unit of General Pathology
Ageing Research Center (CeSI-MeT)
"G. d'Annunzio" University
Via Luigi Polacchi 11, Chieti
and "Mario Sanna" Foundation
Via Emmanueli, 42 – 29121, Piacenza, Italy
Doi:http://dx.doi.org/10.15586/paraganglioma.2019.pr