Pheochromocytoma. Pathophysiology and Clinical Management
Pheochromocytoma
Pathophysiology and Clinical Management

Volume Editor

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Phaeochromocytomas are rare tumours which have nevertheless always caused a huge amount of interest in medical students right up to the most senior endocrinologist. The reasons for this are not difficult to fathom: the tumours form one of the rare causes of hypertension which are almost always curable, and in the majority of cases therapy can cause complete resolution of the high blood pressure. However, in spite of their considerable interest amongst clinicians, it still remains the case that probably the majority of these tumours are only diagnosed post-mortem; in possibly around 50% of patients found with phaeochromocytomas at autopsy, the tumour may have played a direct role in the patient’s demise. There have been vast improvements in the diagnoses of these tumours in recent years in analytical biochemistry and imaging techniques, and we have also become much more aware of the frequency of the genetic causes of phaeochromocytoma and paraganglioma syndromes. This therefore seems to be an apposite moment to review our current knowledge in this area, and accordingly Dr Lehnert has brought together an impressive array of international experts covering all aspects of this fascinating condition. I am delighted to congratulate him on putting together this valuable volume, which I am sure will be of great value to all practising clinicians, and particularly those with interests in cardiovascular medicine, hypertension and endocrinology.

Ashley B. Grossman, London
Preface

This issue in the series *Frontiers of Hormone Research* is devoted to the pathophysiology and clinical management of pheochromocytoma.

Although pheochromocytomas represent rare tumors of the adrenal gland, they are a highly important clinical entity, since they play a distinctive role in the differential diagnosis of hypertension and, in particular, endocrine hypertension. Thus, sound knowledge of the principles of catecholamine metabolism, the clinics and diagnostics of pheochromocytoma are a prerequisite for any physician and scientist interested in hypertension and the adrenal glands.

I am most proud that several renowned scientists and clinicians have devoted state-of-the-art papers to this issue on pheochromocytoma. The principles of catecholamine biosynthesis, metabolism and release are described in greater detail since the biochemical diagnosis of pheochromocytoma is based on the tumor metabolism of catecholamines. These novel findings on catecholamine synthesis and release constitute the basis for diagnostic techniques, e.g. the determination of metanephrines in serum and urine. Although the pathogenesis of pheochromocytoma still remains largely obscure, there are several findings that allow a precise differentiation between benign and malignant tumors and thus further assessment of the prognosis. Within this context it has become clear that approximately one quarter of all presumably sporadic pheochromocytomas harbor a genetic background, e.g. multiple endocrine neoplasia type 2, von-Hippel-Lindau syndrome or familial paraganglioma. Thus genetic testing appears to be mandatory in this tumor. The clinical pictures of sporadic and familial pheochromocytomas, although similar with regard to symptoms due to catecholamine excess, differ with respect to genetic background.
Great advances have been made in the diagnostic imaging of these tumors; scintigraphy with $[^{123}\text{I}]$-metaiodobenzylguanidine is complemented by octreotide scintigraphy in selected cases (i.e. malignant tumors) and, wherever a pheochromocytoma is clinically suspected but cannot be localized by conventional techniques, PET scanning appears to be mandatory. Surgery is still the mainstay of therapy and has evolved into highly differentiated techniques that vary according to the nature of the tumor. If possible, a pheochromocytoma has to be removed by laparoscopic adrenal-sparing surgery. The treatment of malignant pheochromocytoma remains a challenging task and still represents a highly individual approach. The expression of somatostatin receptor subtypes will further allow the application of novel analogs in the future, whereas radiotherapy with metaiodobenzylguanidine and chemotherapy remain ‘the traditional options’.

Hopefully, this book will improve our understanding of the nature of the tumor and thus facilitate the employment of highly effective diagnostic and therapeutic algorithms.

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