

Acknowledgments

In the initial period of my exposure to skull base surgery, my first reference text was *Atlas of Microsurgery of the Lateral Skull Base*. Through this book, I learned about Professor Mario Sanna and Gruppo Otologico and began considering a fellowship at this institute. Professor Sanna welcomed me with open arms at the beginning of March 2010 to begin a year-long fellowship with him. During this time, he has instructed me in detail on the important surgical steps of basic and complex skull base procedures. He has demonstrated precise and fast surgical technique and showed outstanding surgical ability to simplify difficult situations and cases. Also, he has often said to me that a doctor must be frank with his patients, open to his colleagues, and, most of all, honest to himself. He has given me not only the surgical skills but also the philosophy that guides my life as a surgeon and teacher and has gained my undying respect.

After the summer of 2010, he invited me to join him in writing this book and I could feel his enthusiasm for the pursuit of learning and science. He always displayed boundless energy and enthusiasm for scientific writing even after demanding skull base surgeries. His passion motivated me to follow suit and seven months of our work at last bore fruit. I would like to thank him sincerely for giving me a chance to join him in writing this book.

I am especially grateful to Professor Won Sang Lee at the Department of Otorhinolaryngology, Yonsei University, Seoul, Korea, who taught me a great deal on skull base surgery and has encouraged and supported me to complete my fellowship at Gruppo Otologico.

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Finally, I would like to thank my wife, Sea and my young daughters, JiYe and ChaeEun, for enduring the time I spent away from them while completing this book. I also want to express my gratitude to my parents and in-laws for their special support to me and my family.

Seung-Ho Shin, MD

The assessment and management of head and neck paragangliomas provides skull base surgeons with one of their biggest challenges.

The aim of this book is to distil the vast experience of Professor Mario Sanna in the management of these tumors into a comprehensive resource.

It has been an honor to spend a year at the Gruppo Otologico, which has accelerated my development as a skull base surgeon due to the vast volume of cases performed and the unified technique employed.

Involvement in writing this book has also reinforced the importance of a comprehensive knowledge of complex anatomy and radiology, an understanding of the natural history of the underlying disease process, and the need to develop and refine a surgical technique capable of safely addressing all pathologies at the skull base.

Sean Flanagan, MD

Preface

Despite incredible advances in the discipline of skull base surgery, the assessment and management of head and neck paragangliomas remains a demanding task due to the complex anatomical relationship at the skull base. Proper preoperative assessment including detailed neuroradiological investigation is mandatory due to the tendency of these tumors to involve multiple vital structures at the skull base such as the lower cranial nerves and the internal carotid and vertebral arteries. Based on proper preoperative assessment, a detailed, systematic plan of treatment is instituted to maximize tumor resection while minimizing morbidity of the great vessels and lower cranial nerves.

It is our goal in this book to synthesize the current knowledge regarding the pathology and genetics of paragangliomas. Detailed description of the relevant anatomy has always started from a radiological perspective, as it is from this point that a diagnosis is made and subsequent treatment planning is initiated. A surgical framework for the management of these tumors is provided, followed by an extensive section providing a detailed walk-through of specific cases at each stage of disease, highlighting the specific techniques to be used and difficulties expected to be encountered. Each section is illustrated with multiple high-quality surgical and radiological images.

While it is our contention that surgical resection in the hands of highly experienced surgical teams remains the primary management modality in head and neck paragangliomas, knowing when surgery is not indicated is an essential component in the management of these lesions. The accumulated surgical wisdom in this volume has been gained from over 30 years of experience and more than 350 operated cases, and represents a continuing evolution in the optimal surgical management of head and neck paragangliomas.

During this long period we have been exposed to many difficult and complex situations with involvement of the internal carotid and vertebral arteries, recurrent tumors, malignancy, and association with bilateral or multiple tumors; and there were also cases with serious hemodynamic problems.

For these complex paraganglioma cases, we present a detailed decision-making process illustrating the general and specific considerations in planning and successfully executing surgical interventions in a wide variety of challenging clinical scenarios. These include cases of very large tumors with intradural extension or extensive internal carotid involvement, and tumors in patients with complex intracranial arterial and venous circulation. In addition to these scenarios there are also cases of preoperative contralateral lower cranial nerve palsies, patients with recurrent tumors, previously irradiated patients, and patients with malignant tumors.

To reduce the complications resulting from the manipulation of the internal carotid artery, we have introduced stent insertion.

This obviated the need for internal carotid artery occlusion for tympanojugular and vagal paragangliomas and carotid body tumors. Our early results with the use of uncovered stents have shown promising results in terms of carotid artery preservation and gross total tumor removal without serious complications.

Using the Fisch and Mattox classification as a foundation, we subdivide the class A and B tumors into five subclasses (A1, A2, B1, B2, and B3) based on clinical findings. In the A class, the subdivision is based on otoscopic findings and it was formulated after noting that the simple expedient of this examination allowed a more accurate determination of the extent of the disease and subsequently also the nature of the curative surgery required. In the subdivision of the B class, we noted that the extension of the paragangliomas into the hypotympanum and mastoid determined to a large degree the approach required for resection and also the possible postoperative morbidities. In the B3 subdivision, the involvement of the carotid canal in the middle ear was of particular importance as this would necessitate further extension of the approach anteriorly and in some instances procedures to control the carotid were necessary. This modified subclassification is of great assistance in formulating an overall surgical strategy for dealing with these particular tumors.

In this book, we also introduce the new class Vi/e with regard to the involvement of the vertebral artery. Although involvement of the vertebral artery is relatively uncommon, its involvement by tympanojugular paragangliomas can adversely affect the ability to achieve gross total tumor removal. Therefore, this new addition is complementary to the modified Fisch classification and is of great value in management of vertebral artery involvement.

The compilation of this volume was motivated by the desire to encourage and inspire the next generation of skull base surgeons to attain excellence in the management of one of the pinnacles of skull base surgery, in addition to helping refine the skills of those already working in this demanding area.

Acknowledgments

I would like to acknowledge the contribution of my teacher and friend Professor Ugo Fisch of Zurich, Switzerland. His pioneering work and exhaustive study of skull base pathologies resulted in the Fisch classification that has long been widely accepted as a standard classification for skull base paragangliomas. Drawing upon decades of experience and wisdom, he has served as a great teacher, inspiring many with his passion and encouragement. I am therefore honored to be able to introduce a complementary addition to the Fisch classification in this volume, which seeks to extend the scope of knowledge in the area of head and neck paragangliomas.

I am very grateful also to Dr. Paolo Piazza, Interventional Radiologist from the University of Parma for his invaluable work in dealing with the challenging cases we are faced with.

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Mario Sanna, MD