

Preface

In planning this book, I had three major goals. The first was to compile and disseminate all the advances and new information relating to meningiomas which became available in the last 15-20 years. In this time frame, there has been a significant increase in our understanding in regards to the meningioma pathologic classification, the natural history and basic science. Dramatic technological advancements have also been made in diagnostic and interventional radiology as well as in surgical and radiation treatments for meningiomas, such as incorporation of the following in the treatment armamentaria: endoscopy, various skull base techniques, computer-assisted surgery and radiosurgery. Additionally, new information regarding surgical outcome and patient selection for surgery are becoming available, all of which are resulting in a significant change in how neurosurgeons treat patients with meningiomas.

The second goal for this book was to teach and stimulate the next generation of neurosurgeons. Because meningiomas can occur anywhere within the intracranial space or along the spine, for an individual neurosurgeon, mastering the surgery of meningiomas allows one to become a master surgeon. In other words, to learn the basics of meningioma surgery – i.e. surgical decision making, positioning, anatomy, approach, exposure and microdissection, tumor removal, hemostasis, closure- is to learn the basics of neurosurgery. Moreover, meningioma surgery is arguably the most rewarding, challenging and, at times, daunting task for neurosurgeons: rewarding, because of the benign nature of most meningiomas, leading to a possible cure following total removal, challenging because of the tumors' common sites of involvement in proximity to critical neurovascular structures especially when involving the central skull base, and daunting due to the tumor's tendency to recur in higher grades of histologic subtypes and to its frequent involvement of the surrounding skull base bone, dura and neurovascular structures making complete removal often risky or impossible. Although meningioma surgery can be enjoyable and rewarding, I wanted young readers to appreciate and respect the challenging and daunting aspects, which will undoubtedly serve as the stimulus for continued learning, refinement and progress in this field in the future.

The third goal, but the most important one, was to give back to our patients. As neurosurgeons or physicians, we are nothing without our patients. Our patients are truly the backbone of our professional livelihood. It is a great privilege to be able to provide care for other human beings. I firmly believe that the best way to show sincere gratitude to our patients is to not only provide the best care possible, but also to learn from each, so that treatment for the subsequent patients is better and improved.

Editing this book was a much greater task than initially anticipated, with 64 chapters contributed by over 110 distinguished authors from 5 different continents. I am truly honored to be given the opportunity to complete this project which could not have been possible without the support of all the contributors and the publisher, Springer-Verlag. My only regret is that I could not possibly include all of the international experts to join me in this project.

I intentionally solicited differing views and approaches when there are multiple reasonable ways of dealing with the same problem. I thought presentation of multiple ways is superior to pre-selected (and, hence biased) single presentation. Moreover, I wanted very much for young readers to appreciate the fact that there is no single best way of treating certain meningiomas. Hopefully, they will appreciate that whatever technique or approach that results in the best long-term patient outcome in their local setting is what really matters, whether it is surgery vs. radiosurgery for cavernous sinus meningiomas, endoscopic surgery vs. microsurgery in anterior skull base meningiomas, total vs. subtotal resection followed by radiosurgery in parasagittal or skull base meningiomas, anterior vs. posterior transpetrosal approach in petroclival meningiomas, aggressive surgery vs. radiation in optic nerve sheath meningiomas, etc.

This book could not be completed without the valuable assistance of Ms. Christine Moore, an editorial assistant in the Department of Neurosurgery, Cleveland Clinic, and Dr. Burak Sade, my former fellow and present colleague. I cannot thank them enough! I am also greatly indebted to all my mentors: as stated above, they include all my patients, in addition to Professors Eve Marder (neurophysiology professor in college), Alain B. Rossier (college senior thesis preceptor) and John A. Jane, Sr. Lastly, I thank my lovely wife, Heeyang, and my dearest sons, Terry, Nick and Ryan, for their constant support, love and inspiration.

As stated earlier, one of the goals of this book was to teach. However, in completing this book, I became the biggest beneficiary, having learned so much. Just like meningioma surgery, editing this book was immensely enjoyable, rewarding and challenging. If through this book, I have stimulated even a small number of young neurosurgeons to learn and make continued progress in the area of meningiomas, so that they in turn can provide better and improved care for their future patients, I have fulfilled my goals.

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2

Epidemiology

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Introduction

Meningiomas constitute 13–25% of primary intracranial neoplasms.^{1–3} In one population-based study, these tumors accounted for 40% of all primary intracranial neoplasms when tumors diagnosed incidentally at autopsy or by neuroimaging studies were included.⁴ Symptomatic tumors were encountered in 2.0/100,000 of the population and the asymptomatic ones in 5.7/100,000, with an overall incidence of 7.7/100,000. Asymptomatic meningiomas can be found in about 1–2.3% of all autopsies.⁵ However, the classical teaching of the past few decades, which may be erroneous, suggests the most common primary intracranial neoplasms are gliomas (50.3%), followed by meningiomas (20.9%), pituitary adenomas (15%) and nerve sheath tumors (8%).^{6,7} Such discrepancy in these incidence rates underscores the fact that the majority of meningiomas actually remain asymptomatic and undetected during life.⁸ With recent advances in neuroimaging, many asymptomatic meningiomas are being detected today, making the true incidence higher than those previously reported.⁹ It has been shown that the incidence for meningiomas increased 3- to 3.9-fold in the post-computed tomography (CT) decade.^{10,11}

Gender, Race, and Age

Meningiomas show a higher incidence among women as compared to men in most ethnic groups.^{12,13} The male-to-female ratio ranges from 1:1.4 to 2.6.^{14–16} In population-based studies, the mean annual crude incidence rates are reported to be 2–7/100,000 for women and 1–5/100,000 for men.^{1,3}

In a population-based study performed in Los Angeles County, African Americans showed a higher incidence (3.1/100,000) than Caucasians (2.3/100,000).¹² In this study, Asians living in Los Angeles had the lowest rates. In Caucasians, women are approximately twice as likely to develop meningiomas than men, whereas in African American populations the incidence is evenly distributed between males and females.¹ Two studies have also shown a lower incidence of meningiomas

in Asia as compared to western countries,^{17,18} whereas Chi et al. reported no significant difference in the incidence of meningiomas in relation to other intracranial tumors, namely, 20.8% in Korea, 18.5% in Japan, and 16.6% in China.¹⁹ Others have also observed no racial differences in incidence rates of meningiomas.¹³

The mean age at presentation is 56.4 years (range 10–85 years) in males and 55.9 years (range 26–86 years) in females, whereas in the subgroup of malignant and atypical meningiomas, the mean age shifts to 63.2 years (range 51–78 years) in males and 53.6 years (range 28–79 years) in females, a difference that is not statistically significant.²⁰ The incidence of meningiomas increases with age.²¹ In patients older than 70 years, they have been reported as the most common brain tumors, with an incidence of 50.6%. This represents an almost 3.5 times higher incidence in this age group as compared to those under the age of 70, and it applies to both sexes.^{4,22} The age-specific annual incidence rate increases in the eighth decade to 8.4/100,000.¹⁶

Histology and Location

The vast majority of meningiomas (92%) have a benign histology, whereas 8% show atypical or malignant features.²³ The most common histopathological subtype is the meningotheliomatous type (63%), followed by transitional (19%), fibrous (13%), and psammomatous (2%) meningiomas.²⁴ The majority of malignant meningiomas are located over the cerebral convexities. In approximately 73.3–75% of cases these tumors are located in the supratentorial compartment.⁸ The ratio of calvarial to basal skull meningiomas was reported as 2.3:1.²⁵ Extracranial metastases from meningiomas have been considered to be one of the strong indicators of malignancy and have been shown to occur in 11–23% of patients with malignant meningiomas.⁶

The most common locations include parasagittal/falcine 25%, convexity 19%, sphenoid ridge 17%, followed by suprasellar 9%, posterior fossa 8%, olfactory groove 8%, middle

fossa/Meckel's cave 4%, tentorial 3%, peri-torcular 3%, lateral ventricle 1–2%, foramen magnum 1–2%, and orbit/optic nerve sheath 1–2%.¹⁰ Among the parasagittal meningiomas, 49% are located over the anterior one third of the falx, with 29% in the middle third, and 22% along the posterior third.¹⁶ Medial sphenoid ridge meningiomas were more common than middle or lateral sphenoid ridge meningiomas. Multiple meningiomas or meningiomatosis is encountered in 2.5% of meningiomas. The incidence of ectopic location is 0.4% with the vast majority (73%) occurring inside the orbit, paranasal sinuses, eyelids, parotid gland, temporalis muscle, temporal bone, and zygoma. Distant sites have also been reported, such as the lungs, mediastinum, and the adrenal glands.^{6,26}

Recently, Lee et al. demonstrated an association between the histology of the tumor and its site of origin.²⁴ They showed a predominance of meningothelial meningiomas at the midline skull base and spinal locations. Based on this finding, as well as embryological and molecular features, they suggested that this particular subtype of meningiomas may indeed be unique, contrary to the traditional dogma that all benign meningiomas are identical or homogeneous tumors.

Meningiomas in Children

In children, meningiomas account for only 0.4–4.0% of primary intracranial neoplasms.^{1,27–29} The age-adjusted annual incidence was reported to be 1.32/1,000,000.³⁰ There is a male predominance, with a male-to-female ratio of 1.2 to 1.9:1.^{27,30}

The majority of meningiomas in the pediatric age group are located supratentorially (66%), whereas 19% occur in the posterior fossa and 17% present as intraventricular meningiomas.³⁰ They are usually seen in association with neurofibromatosis type 2 (NF-2) or following radiation therapy and show a significantly higher incidence of tumor calcification.^{27,28} In NF-2, it has been estimated that 50% of all patients develop meningiomas, and 30% of these patients have multiple meningiomas.⁶

In children, additional unique features include the significantly increased incidence of atypical (36.4%) and malignant (27.2%) subtypes. In infants, meningiomas are extremely rare and show a higher frequency in males and favor convexity location. On the other hand, there is a smaller incidence of seizure, and dural attachment is less frequently seen on preoperative imaging.¹

Spinal Meningiomas

In women, meningiomas are by far the most common primary spinal tumor, accounting for 58% of all spinal tumors, whereas in men they are third most common primary spinal tumor following gliomas and nerve sheath tumors.³¹ Spinal meningiomas are reported to be more frequent in western countries (25–46%) as compared to Asian countries, for

example, 14.1% in China and 8.6% in Thailand.^{17,19,32} Thoracic spine is the most common site (55–57.1%).^{17,19,32} The male-to-female ratio is 1:4 to 1:5.³³ In females, they are very common in the postmenopausal age group, with the majority (75–87%) occurring over the age of 40.^{34,35}

The tumor is located completely intradurally in 83–90%, extradurally in 5–14%, and both intradurally and extradurally in 5% of the cases.^{32,35} Extradural meningiomas are reported to be more common in children. In 50–68% of the cases, the tumor is located lateral to the spinal cord, in 18–31% posteriorly, and in 15–19% anteriorly.^{32,35}

Histologically, 43.9–56.9% of spinal meningiomas show psammomatous subtype, whereas 28–29.9% are meningothelial, 8–19% transitional, 2.3–5% are fibrous, and 0.6% are malignant meningiomas.^{32,35} The incidence of multiple spinal meningiomas is reported as 1–9%. In a recent review of our series, contrary to the above, meningothelial subtype was the most common (80%) in the spine.²⁴

Radiation

In a population-based study, the incidence of meningiomas among Hiroshima atomic bomb survivors was 8.7/100,000.³⁶ When this population was stratified according to the hypocenter to the explosion, the incidence of meningiomas was much higher in patients who had been closer to the site of explosion. Similarly, the incidence of meningiomas was reported as 9.5/100,000 population in the group who had undergone low-dose radiation treatment as children for tinea capitis in Israel.³⁷

In the medical or occupational setting, no significant associations were observed for diagnostic studies and increased meningioma incidence, but the use of radiation therapy to head and neck for neoplastic conditions has been shown to be associated with an increase in the incidence of meningiomas.³⁸

Radiation-induced meningiomas differ significantly from primary intracranial meningiomas in that their incidence of calvarial location, multiplicity, recurrence rate following complete resection, and malignant histology are higher.²⁵

Occupation

Association of increased risk of meningioma incidence has been suggested for various occupations in the literature. There is a huge diversity in the nature of proposed occupations such as dentists, teachers, managers, social workers, workers in the petroleum, rubber and plastics industry, auto body repairers, painters, chemists, carpenters, cooks, woodworkers, glass-makers, machine operators, as well as military workers, motor vehicle drivers, computer specialists, and so on.^{12,39,40}

Rajamaran et al. have stated that it might be practical to analyze these occupations in two groups.⁴⁰ Groups like teachers, managers, etc., tend to be relatively better educated and would be expected to have a higher awareness of their health status,

which would result in the increased detection of tumors in these groups, by earlier recognition of the symptoms, or by stronger willingness of the individual to seek medical care. On the other hand, exposure to some chemical agents or other environmental factors may be more influential in other groups. In this context, lead, tin, cadmium, benzene, and metal dusts and fumes have been suggested as possible contributors.^{12,39–41}

Cell Phone Use

Over the years, there has been much debate about the potential role of cell phones as a causative factor in the development of brain tumors because of the microwaves emitted by these devices. In a recent population-based case-control study conducted on 366 glioma and 381 meningioma patients with 1494 controls, no association was found between cell phone use and increased meningioma incidence. However, in this study, increased incidence for glioma occurrence was detected in individuals who had used cell phones for more than 10 years.⁴² A similar study in a relatively smaller scale also suggested a low risk for development of high-grade glioma for cell phone users, but the overall results did not show any association for either gliomas or meningiomas.⁴³ The study conducted by the Swedish Interphone Study Group also found no increase in incidence for gliomas or meningiomas located in the temporal or parietal lobes, regardless of tumor histology, phone type, or amount of phone use.⁴⁴

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