The past 30 years have been deemed the golden age of head and neck oncology. With improvements in diagnostic surveillance and therapeutic intervention, advances in patient care have resulted in overall improved survival and quality of life for those affected. Management of tumours of the cranial base, a unique subset of head and neck tumours, has also undergone a renaissance of sorts, with more aggressive resections and expanding multimodality therapy. In this paper, we review existing information concerning the outcomes of management of these tumours and present a rationale for therapy of skull-base tumours.

Surgical therapy of cranial-base tumours is a relatively new concept. Just as neurosurgeons once deemed intracranial tumours with cranial-base involvement unresectable, head and neck surgeons originally had little success with these types of lesions, feeling classical approaches (such as lateral rhinotomy) precluded safe and complete resection of masses breeching the skull base. A unimodal approach was recognized as insufficient for surgical management of these tumours. As surgical oncology progressed with the concept of en bloc resection, the desire of surgeons to resect complete anatomical units resulted in the development of novel approaches to cranial-base tumours. Dandy first published his experience with a combined intracranial and extracranial approach in 1941 in his work with orbital tumours. This combined approach gained popularity and was adopted by head and neck surgeons, via Ketcham et al in 1963, for the resection of paranasal sinus malignancies via anterior craniofacial resections.

The advent of the combined approach to cranial-base surgery stemmed from a collaboration between head and neck surgeons and neurosurgeons. Since then, the range of diagnostic and therapeutic options available has expanded and the interdisciplinary relationship has grown to involve an increasing number of physician groups as well as ancillary personnel; each group plays a key role in the care of these patients. The team approach is essential to proper care and should involve head and neck surgeons, neurosurgeons, intraoperative pathologists, plastic and reconstructive surgeons, medical and radiation oncologists, neuroradiologists, speech pathologists, physical and occupational therapists, and prosthetists. With these global advances, surgeons can perform more complex procedures for these patients, but the question as to whether these advances improve patient outcome arises.

The data for cranial-base surgery are limited and have generally been presented as case studies or retrospective analyses. Prospective clinical trials to investigate important issues, especially outcome measures, have not been performed and would be extremely difficult due to the limited experience in most centres, tumour variables (pathological type, grade and stage), and patient variables (general condition, prior disease, surgery or radiation), as well as the difficulty of standardizing treatment algorithms for these various diseases. There is also a lack of uniformity among investigators in the reporting of the specifics of these patients and their tumours. In general, an analysis of our current rationale for therapy is based on published retrospective studies that are somewhat limited and difficult to compare and extrapolate.

The historical aspect of cranial-base surgery was reviewed by O'Malley and Janecka, who described a progression through several periods, each of which has seen surgical advances and increased survival. The first era, from the 1960s to the 1970s, can be characterized by a high complication rate and poor survival despite large selection bias because many patients were deemed inoperable due to extensive local involvement. The authors of studies reviewed for this time period did not operate on certain tumours such as those extending into the
pterygopalatine fossa or intracranially. The second era, from the 1970s to the 1980s, showed similar complication and survival rates, despite the more extensive procedures performed. More aggressive techniques allowed the removal of infratemporal and pterygopalatine fossa tumours as well as neoplasms extending intracranially and even intraparenchymally. During the third period of skull-base surgery (1980s and beyond), these extensive procedures continued to be performed with improved local control, a significant decrease in complication rates, and an improvement in survival (Table). However, this analysis did not address functional status as an outcome measure.

Janecka et al published results from 183 patients who underwent surgery for cranial-base tumours, 67% of which were malignant (most commonly squamous cell carcinoma and adenoid cystic carcinoma). From a surgical point of view, negative margins were achieved in 44% of patients. Oncological results showed a 30-month survival of 67% with no evidence of disease in 63%. The overall complication rate was 33% (21% surgical, 12% medical). Overall functional status, as measured by the Karnofsky scale, was improved: 39% of patients were improved, 44% were unchanged, and 15% scored lower postoperatively.

Another outcome analysis, published in 1994 by Irish et al, concerned 77 cases of skull-base tumours. In an effort to organize their results and to improve standardization of reporting for cranial-base tumours, a staging system based on anatomical location was introduced. The delineation of these regions (I, II and III) are based on the portion of the skull base involved and also correlates somewhat with the approach needed for resection. Region I involves the anterior cranial fossa and the clivus; most of these tumours originate in the paranasal sinuses or intracranially, and are approached via craniofacial resection. Region II tumours involve the middle cranial fossa, generally via direct extension from pterygopalatine or infratemporal fossa tumours through the foramina. Region III tumours originate in or near the temporal bone and extend into the middle or posterior cranial fossa. In general, region II and III tumours are surgically approached using a more lateral procedure, such as an infratemporal approach, facial translocation or a variant of temporal-bone resection.

When surgical outcomes were analysed for each anatomical subunit, patients with region II tumours had very poor survival, 45% at 2 years and 0% at 4 years (n = 71), despite a lower malignancy rate (71%) than the other regions (I = 76%; III = 88%). The overall 2- and 4-year survival rates were 80% and 71% (n = 77), which shows that patients with pterygopalatine and infratemporal fossa tumours involving the skull base have a very poor prognosis despite aggressive surgical resection. The overall complication rate in this study was 44% and increased dramatically to 64% in cases with tumours involving more than one region and requiring more extensive resection. Despite its limited size, this study infers a reanalysis of the management of certain cranial-base tumours, especially extensive, region II tumours.

Shah et al reported their experience with craniofacial resection for anterior cranial-base tumours in 1992 (n = 71). Remarkable results include no difference in survival between previously treated and untreated patients. Also, local control was achieved in 71% of patients with negative margins and 67% of patients with positive margins. O'Malley and Janecka have also reported no statistically significant difference in survival between patients with positive and negative margins. This underscores the importance of the surgeon's interpretation and use of pathology in these cases. In general, it is extremely difficult to fully and properly inspect the entire three-dimensional margins of such a large en bloc resection. Intraoperative consultation with a pathologist is important in order to properly orient the specimen and to inform the pathologist of pertinent patient and surgical details. Survival rates for tumours involving the dura were not statistically different from those involving the brain parenchyma; however, these patients fared much worse than those without penetration of the dura (p = 0.02) in terms of survival.

Despite the surgeon's expanding armamentarium, one must focus on endpoint analysis. First, the detection of both primary and recurrent neoplasms has become much more sensitive and offers more information concerning surrounding structures. Although data may be somewhat obscured at some tissue interfaces, bony invasion is more reliably detected with advances in computed tomography. Magnetic resonance imaging allows determination of the degree and extent of dural and intraparenchymal involvement. Image-guided surgery uses both of these resources and can improve orientation and familiarization with the surgical landscape. Also, positron emission tomography shows promise for the early detection of
recurring disease. Arteriography, carotid artery balloon occlusion and xenon blood-flow studies are now available to help estimate the results of carotid embarrassment or the need for carotid bypass. Advanced techniques in endoscopic sinus surgery allow less invasive approaches to craniofacial resections and can preclude the need for facial incisions. Also, advances in technique and instrumentation for microsurgery (especially involving the cavernous sinus), brachytherapy, nerve grafting and radiotherapy (including stereotactic radiation) offer more diverse treatment options, but the impact of each is difficult to isolate at this time.

Perhaps the most important issue that has affected cranial-base surgery deals with the advancements in reconstruction of these potentially large en bloc resections. The modernization of techniques for local fascial flap (especially the temporoparietal fascial flap) and myocutaneous pedicled flap, and the development of the microvascular free flap allow more appropriate and viable reconstructions and help separate intracranial and extracranial contents. Several authors have postulated that these advances in separation (especially when dealing with craniofacial resections and the nasal cavity) are responsible for the decreased complication rate in the third era of cranial-base surgery.

In the past 15 years, we have managed more than 3,100 head and neck malignancies. Those invading the infratemporal skull base, sphenoid sinus, clivus and brain have been universally fatal by 5 years. Multimodal therapy (surgery, radiation and chemotherapy) has not improved the prognosis for these tumours. From our experience and the published literature, management of malignancies of the head and neck that invade the infratemporal skull base has involved various radiotherapeutic modalities over the past decade due to the extremely poor prognosis for these lesions. For malignant skull-base tumours, contraindications for resection at our institution include involvement of brain parenchyma and invasion of the walls of the sphenoid sinus (except the anterior wall), the clivus, the intrapetrous carotid, the optic chiasm or the infratemporal skull base. Piecemeal removal of malignant lesions with microscopic close margins can only be condemned. Although the pathologist's role is pivotal in this situation, only the surgeon knows the adequacy of his or her resection.

When dealing with benign skull-base tumours, the extent of resection must be balanced against iatrogenic neurological compromise, with care to leave the patient with an acceptable functional status. We have performed 107 craniofacial resections over this period with similar results to those described in the literature, except for patients with sinonasal undifferentiated carcinoma, where the success has been dismal. Extensive discussion with the patient is crucial, and the following are used as contraindications for resection of benign tumours at our institution: extensive involvement of both cavernous sinuses where significant neurovascular injury would occur, involvement of the brainstem with pontine blood supply to the tumour that cannot be separated from the pons, and involvement of the optic chiasm where the tumour cannot be separated without sacrificing both optic nerves. The following are not advised: resection of sufficient craniofacial structures to make reconstruction unacceptable, resection involving bilateral long-tract cranial nerves, and resection of sufficient brain parenchyma (especially the dominant temporal lobe) that may make the patient severely debilitated.

In conclusion, our rationale for therapy of cranial-base tumours is individually tailored by a team of physicians and somewhat molded by the patient's wishes. It is of utmost importance that patients and their families have an understanding of what morbidity is anticipated and the extent of the neurological compromise. Although surgeons have an increasing ability to perform and reconstruct extensive en bloc resections of cranial-base tumours, it is questionable whether these procedures improve patient outcome in all instances. The surgeon and the patient, as leaders of the therapeutic team, are best prepared to develop their rationale for therapy armed with the knowledge of existing outcome analyses tailored to each patient's specific case.

References

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