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Orin Bloch and Franco DeMonte

## **Management Considerations for Malignant Tumors of the Skull Base** **1**

Franco DeMonte

The last two decades have brought refinements in diagnostic imaging, instrumentation, and microvascular reconstruction, and an improved overall appreciation of the anatomy of the skull base, both open and endoscopic. These refinements have extended the boundaries of tumor resection and have obviated adjuvant therapies in some patients with benign or low-grade tumors. In patients with high-grade malignancies, however, a carefully constructed multimodal treatment plan, incorporating surgery, radiation therapy, and chemotherapy, is necessary to maximize patients' outcomes.

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Joshua Marcus, Ilya Laufer, Babak Mehrara, Dennis Kraus, Bhuvanesh Singh, and Mark H. Bilsky

Combined anterior cranial base resection is the mainstay of therapy for skull base malignancies. Improvements in surgical techniques and reconstruction have led to a reduction in morbidity and overall better survival rates. Meticulous attention to dural and skull base reconstruction is essential for reducing the major complications, including cerebrospinal fluid leak and pneumocephalus. Complications can be devastating, but timely effective management can limit the severity.

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Michael E. Ivan, Arman Jahangiri, Ivan H. El-Sayed, and Manish K. Aghi

The use of minimally invasive approaches to the anterior skull base is a valuable tool to improving the treatment in patients with aggressive anterior skull base neoplasms. This article discusses the history, advantages and disadvantages relative to open approaches, the corridors and pathways used in approach, the equipment and operating room setup, perioperative care, and complication avoidance. Although outcomes are difficult to compare to open approaches, due to often small and varying patient cohorts, these approaches continue to gain acceptance as an effective treatment of anterior skull base tumors in the experienced surgeon's hands with proper patient selection.

## **Sinonasal Carcinomas: Epidemiology, Pathology, and Management** **39**

Stephan K. Haerle, Patrick J. Gullane, Ian J. Witterick, Christian Zweifel, and Fred Gentili

Sinonasal carcinomas are uncommon neoplasms accounting for approximately 3% to 5% of all upper respiratory tract malignancies. Sinonasal malignancies in most cases do not cause early symptoms and present in an advanced stage of disease. Exact staging necessitates a clinical and endoscopic examination with biopsy and imaging. Tumor resection using an open or endoscopic approach is usually considered the first treatment option. In general, sinonasal carcinomas are radiosensitive, so adjuvant or neoadjuvant radiation treatment may be indicated in advanced

disease. Multidisciplinary surgical and medical oncologic approaches, including ablation and reconstruction, have enhanced the survival outcome over the past few decades.

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Thomas J. Ow, Diana Bell, Michael E. Kupferman, Franco DeMonte, and Ehab Y. Hanna

Esthesioneuroblastoma is a rare malignant neoplasm in the olfactory region of the nasal cavity and anterior skull base. Diagnosis and staging require anatomic imaging and careful pathologic assessment. Standard treatment is anterior craniofacial resection with postoperative irradiation. The role for chemotherapy is not defined, but is generally for the most advanced cases and used in the neoadjuvant setting and/or postoperatively with irradiation. Prognosis is favorable; however, metastasis rates remain relatively high. Regional and distant metastasis portends a poor outcome. Intensity-modulated radiation treatment and endoscopic surgery have reduced morbidity, but outcomes with these techniques must be fully evaluated.

**Head and Neck Sarcomas: Epidemiology, Pathology, and Management** 67

James Paul O'Neill, Mark H. Bilsky, and Dennis Kraus

Sarcomas of the head, neck, and skull base represent a heterogeneous group of tumors with distinct prognostic features. There have been significant improvements in characterizing these sarcomas using traditional morphologic assessments and more recent immunohistochemical analysis. Surgery is the mainstay of treatment followed by radiation therapy. Treatment modalities have changed in select pediatric sarcomas, for which new chemotherapeutic combinations have improved survival statistics. The high rate of distant failure emphasizes the need for novel systemic and directed molecular therapies. Tumor grade, size, and margin status are key factors in survival.

**Skull Base Chordomas: Clinical Features, Prognostic Factors, and Therapeutics** 79

Arman Jahangiri, Brian Jian, Liane Miller, Ivan H. El-Sayed, and Manish K. Aghi

Chordomas of the skull base are one of the rarest intracranial malignancies that arise from ectopic remnants of embryonal notochord. The proximity of many chordomas to neurovascular structures makes gross total resection difficult, and the tendency for recurrence leads to the routine use of adjuvant postoperative radiation. Several surgical approaches are used ranging from extensive craniotomies to minimally invasive endonasal endoscopic approaches. In this review, the histopathology and epidemiology, imaging characteristics, surgical approaches, adjuvant therapies, prognostic factors, and molecular biology of chordomas are described.

**Skull Base Chondrosarcoma: Evidence-Based Treatment Paradigms** 89

Orin Bloch and Andrew T. Parsa

Chondrosarcomas are indolent but invasive chondroid malignancies that can form in the skull base. Standard management of chondrosarcoma involves surgical resection and adjuvant radiation therapy. This review evaluates evidence from the literature to assess the importance of the surgical approach and extent of resection on outcomes for patients with skull base chondrosarcoma. Also evaluated is the ability of the multiple modalities of radiation therapy, such as conventional fractionated radiotherapy, proton beam, and stereotactic radiosurgery, to control tumor growth.

Finally, emerging therapies for the treatment of skull-base chondrosarcoma are discussed.

## **Temporal Bone Malignancies**

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Paul W. Gidley and Franco DeMonte

Primary temporal bone tumors are rare. Suspicious lesions of the ear canal should be biopsied for diagnosis. Surgical resection to achieve negative margins is the mainstay of treatment. Small tumors can be treated with lateral temporal bone resection. Parotidectomy and neck dissection are added for disease extension and proper staging. Higher staged tumors generally require subtotal temporal bone resection or total temporal bone resection. Adjuvant postoperative radiotherapy has shown improved survival for some patients. Chemotherapy has an emerging role for advanced stage disease. Evaluation and management by a multidisciplinary team are the best approach for patients with these tumors.

## **Craniofacial Reconstruction Following Oncologic Resection**

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Matthew M. Hanasono and Theresa M. Hofstede

The ability to reliably reconstruct complex and sizable wounds has decreased the morbidity of skull base surgery substantially, preventing major complications and allowing treatment of tumors previously considered inoperable. Addressing facial nerve function with static and dynamic procedures as well as fabrication of craniofacial prostheses to replace delicate facial landmarks has further increased surgeons' ability to restore the appearance and function of the face.

## **Radiotherapy for Malignant Tumors of the Skull Base**

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Julian Johnson and Igor J. Barani

Malignant tumors of the skull base are a fascinating group of tumors arising via disparate causes leading often to similar presentations. This article explores radiotherapy techniques applied to this group of malignancies, with a focus on providing general overview and guiding readers to primary sources to achieve greater depth. The outcomes and effects of radiation, therapeutic radiation modalities and delivery system are discussed. Equipped with these basic principles, practitioners will have general guidance for rational treatment modality selection for patients with skull base tumors.

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