A Concise Review of Irradiation for Temporal Bone Chemodectomas (TBC)

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Abstract

Chemodectomas of head and neck region are rare, highly vascularized tumors which are categorized with respect to their site of origin and may be observed with periodical imaging or treated using either a single modality or multimodal approach including the primary management strategies of surgery and Radiation Therapy (RT). Temporal Bone Chemodectomas (TBC) include the tympanomastoid chemodectomas with modified Fisch Class A and B, and tympanojugular chemodectomas with modified Fisch Class C and D. An indolent disease course with low growth rate is typical for the majority of these mostly benign tumors, however, abrupt manifestation with severe symptomatology may also occur rarely in affected patients due to the mass effect with or without local invasion of critical neurovascular structures such as the internal carotid artery, jugular bulb, and lower cranial nerves. Affected patients may suffer from a plethora of symptoms such as otalgia, vertigo, pulsatile tinnitus, hearing impairment, headache, dysphagia, and dizziness. While surgery has been the traditional primary mode of treatment for these tumors, RT and more recently radiosurgery have been incorporated in management to achieve optimal therapeutic outcomes. Herein, we provide a concise review of RT for temporal bone chemodectomas.

Introduction

Chemodectomas of head and neck region are rare, highly vascularized tumors which are categorized with respect to their site of origin and may be observed with periodical imaging or treated using either a single modality or multimodal approach with the primary management strategies of surgery and Radiation Therapy (RT) [1–6]. Temporal Bone Chemodectomas (TBC) include the tympanomastoid chemodectomas with modified Fisch Class A and B, and tympanojugular chemodectomas with modified Fisch Class C and D [6,7]. First description of TBC dates back to 1945 [8]. An indolent disease course with low growth rate is typical for the majority of these mostly benign tumors, however, abrupt manifestation with severe symptomatology may also occur rarely in affected patients due to the mass effect with or without local invasion of critical neurovascular structures such as the inner ear, internal carotid artery, jugular bulb, and lower cranial nerves [1,2]. Affected patients may suffer from a plethora of symptoms such as otalgia, vertigo, pulsatile tinnitus, hearing impairment, headache, dysphagia, and dizziness [1,2]. While surgery has been the traditional primary mode of treatment for these tumors, RT and more recently radiosurgery have been incorporated in management to achieve optimal therapeutic outcomes [1–7]. Herein, we provide a concise review of RT for temporal bone chemodectomas.

Irradiation for management of temporal bone chemodectomas

A growing body of literature spanning about three-quarters of a century has focused on different aspects of chemodectomas and their management with surgery, RT, and radiosurgery [1–76]. Over the years, there have been significant improvements in the discipline of radiation oncology incorporating contemporary radiotherapeutic strategies such as Image Guided Radiation Therapy (IGRT), Adaptive Radiation Therapy (ART), Intensity Modulated Radiation Therapy (IMRT), Breathing Adapted Radiation Therapy (BART), and stereotactic irradiation with Stereotactic Radiosurgery (SRS), Hypofractionated Stereotactic Radiation Therapy (HFSRT), and Stereotactic Body Radiation Therapy (SBRT) [1,2,77–83]. Radiosurgery in the forms of SRS...
and HFSRT has proved to be safe and effective for management of several intracranial and extracranial disorders [1,2,84-102].

While studies conducted in the earlier periods may be subject to considerations about the use of relatively older RT techniques to deliver a wide range of doses for management, current RT practice involves incorporation of modernized treatment equipment and planning systems along with optimization of delivered dose under image guidance for an improved therapeutic ratio for TBC. Prescribed doses for either conventionally fractionated RT or radiosurgery protocols are being substantiated with accumulating evidence. Normal tissue sparing has been improved by use of intensity modulation techniques. In this context, conventionally fractionated RT and radiosurgery as SRS, FSRT, or HFSRT are now considered as a viable part of multimodality TBC management. While management of advanced, incompletely resected, or recurrent lesions with irradiation has been widely accepted as common practice, accumulating evidence in favor of irradiation has been supported by several studies as well as systemic reviews and metaanalyses assessing its utility as the primary mode of management [56,63-65,73-76,103-110].

In the context of irradiation options, radiosurgery offers the advantage of delivering stereotactically focused and high dose radiation to the well defined treatment volume with steeper dose gradients around the target thereby achieving improved normal tissue sparing. Ultimate effect is possibly achieved by complex interactions of endothelial inflammation, vascular endothelial damage around the tumor-supplying vessels and apoptosis [111,112]. Irradiation of small volumes typically without any margins allows for delivery of high doses with radiosurgery. Clearly, this highly sophisticated technology warrants robust immobilization and image guidance for precise radiation delivery. Negligible doses around the target with radiosurgery allows for respecting the tolerance of critical surrounding structures, leading to an improved toxicity profile compared to surgery [103,104]. Additional favorable aspects of radiosurgical management may include shortening of overall treatment time with expedited recovery typically as an outpatient procedure, earlier return to daily life, improved functional preservation and quality of life, and cost-effectiveness [1,103-106].

In a comprehensive review of paraganglioma management using irradiation and surgery, local control rates were found to be 93.7%, 89.1%, and 78.2% with treatment modalities of SRS, external beam RT, and surgery, respectively with statistical significance [65]. Mortality rate was 9.2% with surgery vs none with SRS [65]. As for morbidity, cranial nerve palsies were increased with surgery but decreased with external beam RT and SRS compared to the preoperative levels [65].

Clearly, comparative assessment of treatment modalities based on retrospective data may be prone to confounding factors such as considerable variations in patient and tumor characteristics among the included studies. Nevertheless, accumulating evidence suggests that RT and particularly radiosurgery is being increasingly considered to have an expanding indication for management of these tumors.

**Conclusion and future perspectives**

Chemodectomas of the head and neck region require thorough consideration since management with a given modality may be associated with substantial deterioration in quality of life of the affected patients along with morbidity and even mortality due to the intricate anatomy and intimate association of some lesions with vital neurovascular structures. There has been significant progress from the standpoint of both surgery and RT in recent years with particularly radiosurgical management being supported by accumulating data and suggested as a viable alternative or supplementary treatment modality for a considerable group of affected patients. Nevertheless, rather than selecting one modality over another, it is more plausible to individualize management taking into account factors including age, tumor size, location, growth rate, symptomatology, comorbidities, performance status and patient preferences to improve the therapeutic ratio. Particle therapy, staged treatment of selected patients, and incorporation of systemic agents merit further research to improve outcomes of management.

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