

Surgical Resection of a Small Cell Carcinoma Primary Tumor of the Parotid Gland with Perineural and Skull Base Involvement

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Abstract

Background: One of the rarest tumors of the parotid gland is a Small Cell Carcinoma (SmCC) of neuroendocrine or ductal origin. These lesions tend to be aggressive, often invading bone and nerve and carry a grave prognosis for the patient. Presently there is no standardized treatment protocol and no consensus to tumor management.

Case Report: This case report is of a 70 year-old male with a right-sided facial mass, facial pain, severe otalgia, weight loss, and right-sided facial paralysis. Subsequent imaging and Fine Needle Aspiration (FNA) revealed T4aN2bM0 SmCC of the parotid gland with facial nerve enhancement and skull base erosion. The patient underwent major extirpative surgery including radical neck dissection, total parotidectomy with facial nerve resection and cable graft, and partial temporal bone resection as the initial step in a combined modality therapy. The patient has done very well post-operatively.

Conclusion: This case report describes the successful, aggressive surgical management of locally and regionally advanced SmCC of the parotid gland. Although the prognosis for patients with SmCC of the parotid gland is typically grave, the absence of distant metastatic disease in the presented case provided the rationale for an aggressive surgical approach with a curative, rather than palliative, focus.

Keywords: Parotid; Small cell carcinoma; Combined modality; Surgery; Radiation; Aggressive

Introduction

Salivary gland malignancies are most commonly found in the parotid gland. Neuroendocrine Carcinoma (NEC) is a rare form of salivary gland cancer, presenting as one of three subtypes: atypical carcinoid, moderately differentiated carcinoid, and small cell carcinoma (SmCC) as per WHO designations, and NEC I, II, and III from the Wick Classifications [1-3]. Of these, SmCC has the worst prognosis with 2 and 5 year survival rates of 70 and 46%, respectively. Typically, SmCC metastasizes hematogenously, rather than via lymphatic routes [4]. However, as evidenced in the following case, when the tumor does metastasize through the lymphatic system, it is very destructive.

The location of the parotid gland relative to the facial nerve, stylomastoid foramen, and skull base presents therapeutic challenges to disease management. There is no universally accepted “gold standard” approach to management of primary SmCC of the parotid gland due to a scarcity of reported cases [4]. Among these are a number of reports that describe successful management with either surgery or radiation alone [5-6]. The following case is unique with respect to histology as well as its aggressive pattern of local and regional involvement, thus posing a particular challenge to clinical management.

Case Report

A 70 year-old male presented to our practice with a 2 month history of right facial mass, and complaints of facial paralysis, numbness around the lips, and progressive pain localized to the face and teeth. Fine Needle Aspiration (FNA) revealed SmCC with sheets of dyscohesive small cells with hyperchromatic nuclei, a finely dispersed chromatin pattern, nuclear molding, minimal cytoplasm, numerous mitotic figures, apoptotic bodies, and necrosis. Immunohistochemical stains demonstrated keratin positivity (CAM5.2) with a dot like perinuclear pattern. Stains for neuroendocrine markers (synaptophysin and CD56) were also positive (Figure 1). CT of the neck and skull base MRI and CT revealed a 5.9 cm × 4.9 cm × 6.1 cm mass encasing the right styloid

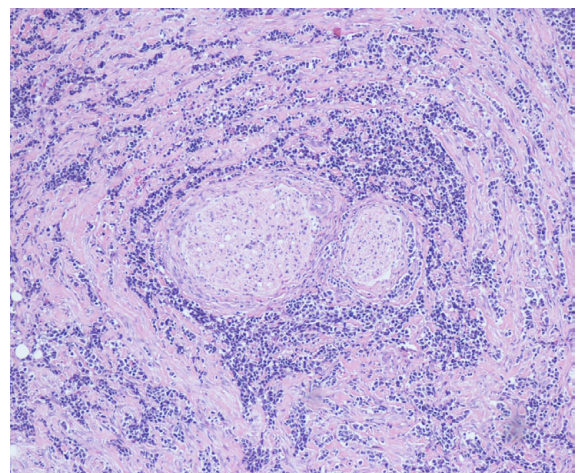


Figure 1: Small cell carcinoma demonstrating perineural invasion. Hematoxylin and eosin stain. 100X magnification.

process and less than 180 degrees of the external carotid artery. The mass was also shown to compress the right internal jugular vein all the way to the stylomastoid foramen (Figures 2 and 3). Bony erosions and

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Figure 2: Axial CT of the skull base shows right parotid mass invading the stylomastoid foramen.

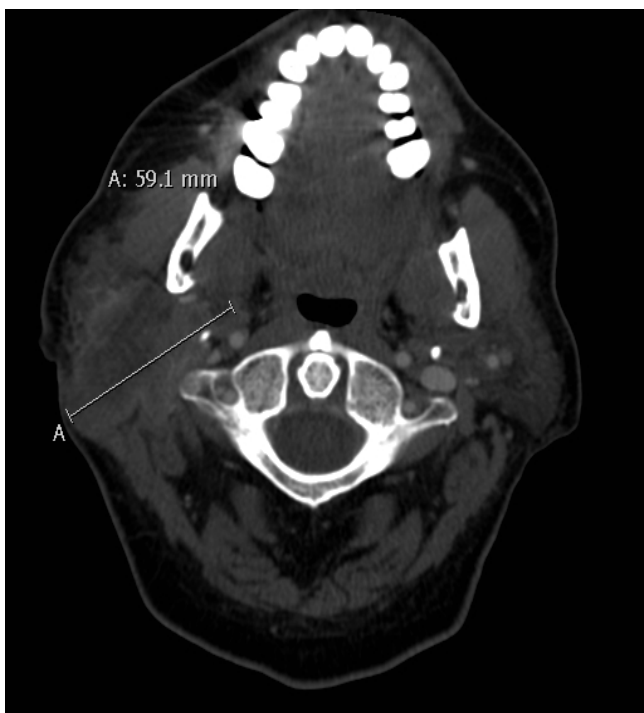


Figure 3: Axial CT of parotid tail and right parotid mass at the level of C2 with an inferior diameter of 59.1 mm.

suspected perineural spread were evident, along with enlarged level II lymph nodes.

A PET scan was obtained revealing increased activity in the primary

site and ipsilateral level II nodes, with no evidence of distant metastases. During the process of this evaluation and subsequent consultation with radiation and medical oncology, the patient developed progressive issues with pain and nausea, requiring a hospitalization. Given the severity of the symptoms, coupled with the localized nature of the disease, a decision was made to proceed with aggressive surgical intervention, to be followed with chemotherapy and radiation.

The surgical procedure involved partial temporal bone resection to address skull base as well as facial nerve involvement at the stylomastoid foramen, and to control the internal jugular vein above the area of involvement. Excision of a large area of postauricular skin was necessary due to dermal invasion, but the pinna was spared. Total parotidectomy with sacrifice of the facial nerve and the external carotid artery was completed in conjunction with a radical neck dissection. The facial nerve was cable grafted using the contralateral greater auricular nerve which was sutured into place using microsurgical technique between the intertemporal stump of the facial nerve and the distal stump of the buccal branch. The temporal bone defect was obliterated with fat and a temporalis muscle rotation flap. Reconstruction of the skin defect was accomplished with a cervicofacial rotation flap.

The en bloc resection specimen revealed a neuroendocrine carcinoma with replacement of parotid tissue and invasion of cartilage, subcutaneous tissue, and dermis. 11 of 23 lymph nodes were positive. Lymphovascular and perineural invasion were both present along with extra nodal extension. Following surgery, the patient received induction chemotherapy with cisplatin and VP-16, followed by radiation therapy, and experienced a marked improvement in pain and functional capacity. Attempts at palliation of facial nerve dysfunction included gold weight placement in the upper eye lid, to assist with eyelid closure, and lower lid tightening. There has been return of some resting muscle tone to the mid face.

Discussion

While SmCC of the salivary glands affords a better prognosis than pulmonary and other head and neck SmCC, it can still be locally aggressive, with a high metastatic potential [7]. Lymphatic and distant spread can occur in as many as 47% and 67% of patients, respectively [8]. Two year survival rates of 38 to 70% and five year survival rates of 13 to 46% have been reported [8,9] Negative prognostic factors include a tumor greater than 3 cm, negative immunostain for cytokeratin 20, and dermal invasion [8]. Although Merkel cell carcinoma cannot be distinguished from SmCC by morphologic and immunophenotypic characteristics alone (including a majority of both with CK20 perinuclear dot like positivity), the clinical presentation in this case was a primary salivary gland tumor rather than a primary cutaneous malignancy. CD20 (a B cell lymphoid marker) was negative, which is not really relevant to the diagnosis. The authors recognize that although a specific diagnosis can only be made by FNA in approximately 60-75% of malignant salivary gland tumors, in this case the morphology (mitotic activity, apoptosis, necrosis, etc.) and immunohistochemical features characteristic of a high grade neuroendocrine carcinoma were present. Additionally, the availability of an adequate cell block from which immunohistochemistry could be performed allowed for a specific diagnosis.

The presence of facial nerve palsy has been associated with as high as ten-fold decreases in survival. Even in cases of parotid malignancy with negative surgical margins, the local recurrence rate is as high as 17% [10]. Lateral temporal bone resection may produce the greatest decreases in mortality for high stage malignancies. Aggressive resection

of skull base and temporal bone tumors does afford a survival benefit to patients that local radiation and chemotherapy cannot accomplish; however, this management strategy has drawbacks. A retrospective analysis of 43 cases of T1/T2 external auditory canal carcinomas of the lateral temporal bone revealed that lateral temporal bone resection (LTBR) decreases mortality but increases morbidity as a result of facial nerve paralysis and conductive hearing deficits, compared to local canal resection (LCR) [11].

The LTBR performed in this case required facial nerve sacrifice and cable nerve grafting, as well as requiring other interventions for improving on postoperative quality of life. Past investigations have downplayed the contributions of physical disfigurement, conductive hearing loss and facial nerve dysfunction to postoperative quality of life in patients undergoing LTBR for parotid and other temporal bone malignancies [12,13]. Our patient experienced a conductive hearing deficit similar to that observed with LTBR treatment of external auditory carcinoma.

The gold weight placement and lower lid tightening to enhance eyelid closure is a reversible procedure that can be readily undone should facial nerve function return. When indicated these procedures provide an immediate aesthetic and functional solution to orbital branch symptoms. In a prospective study with 30 patients, gold weight placement in the upper eye lid and lower lid tightening achieved favorable outcomes while maintaining the option to reversed [14].

The great auricular nerve and sural nerve are common structures used for cable nerve grafting of the facial nerve due to the limited functional deficits from their harvest. Great auricular and sural nerve interpositioning and grafting can accomplish up to a House-Brackmann level three facial nerve function; however, successfully restoring function of the facial nerve with grafts is limited by the duration of facial paralysis of 5 months or less, according to one study [15,16]. Our patient underwent facial nerve autograft using the contralateral greater auricular nerve as a graft between the distal and proximal stumps of the facial nerve. At present, the restoration of facial nerve function is limited to return of some resting muscle tone in the mid face.

At the time of this writing, approximately 16 months post treatment, the patient remains free of disease and reports exceptionally high function and quality of life. Following his treatment for SmCC of the parotid gland he reports resuming an active lifestyle, frequently participating in triathlons.

Conclusions

This case report describes extremely favorable outcomes resulting from an aggressive surgical approach to locally and regionally advanced SmCC of the parotid gland. Following surgery, the patient recovered rapidly, demonstrating significant improvement in his presenting symptoms. This allowed for timely initiation of adjuvant chemo and radiation therapy. Although long-term prognosis is yet to be determined, the palliative value of this approach seems clear. Surgical intervention should be considered as a component of a multidisciplinary approach to select cases of advanced SmCC of the parotid gland, particularly in cases without distant metastatic spread.

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