Acinic Cell Carcinoma of the Parotid Gland: A Case Report and Review of the Literature

Ilson Sepúlveda\textsuperscript{a, d}, Michael Frelinghuysen\textsuperscript{b}, Enrique Platin\textsuperscript{a, d}, M. Loreto Spencer\textsuperscript{c}, Alexis Urra\textsuperscript{a} Pablo Ortega\textsuperscript{a}

\textsuperscript{a}ENT-Head and Neck Surgery Service, General Hospital of Concepcion, \textsuperscript{b}Department of Radiation Oncology, Oncology Service, General Hospital of Concepcion, and \textsuperscript{c}Department of Pathology, General Hospital of Concepcion, University of Concepcion School of Medicine, Concepcion, and \textsuperscript{d}School of Dentistry, Finis Terrae University, Santiago, Chile; \textsuperscript{e}School of Dentistry, University of North Carolina, Chapel Hill, N.C., USA

Key Words
Computed tomography · Magnetic resonance imaging · Parotid gland · Parapharyngeal space · Tumor

Abstract
We report on a patient who was referred to the ENT service following an incidental finding on an MRI scan of the brain. It revealed a mass in the right parapharyngeal space, and additional imaging confirmed the presence of a solid cystic expansive mass with moderate enhancement following contrast media injection. The patient was treated with a total parotidectomy followed by radiotherapy. Currently, the patient is disease-free without any complications.

Introduction
Acinic cell carcinomas (ACCs) are uncommon salivary gland tumors that most commonly occur during the fifth and sixth decades of life. The incidence of ACCs is slightly higher in women than in men. There are no clear characteristics of ACCs found on CT, MRI and ultrasound imaging. The most common finding is a well-defined solid cystic mass. The preferred treatment is complete surgical excision.
Case Report

A 62-year-old female was referred to the ENT service as a result of an incidental finding on an MRI scan of the brain. The MRI revealed a mass in the right parapharyngeal space. Her clinical history included trigeminal neuralgia and a normal laryngoscopy. CT and MRI studies were performed to identify the nature of the mass.

The CT revealed an iso-hypodense solid cystic expansive mass in the right parapharyngeal space, measuring 3 cm in diameter. The mass was well defined, showing moderate enhancement following intravenous contrast injection (fig. 1). It displaced the anterior and medial internal carotid arteries and the internal jugular vein (fig. 2). Lymphadenopathy was present in the carotid and bilateral submandibular spaces, measuring up to 14 mm in diameter.

The MRI revealed a solid cystic expansive mass in the right parapharyngeal space arising from the deep parotid lobe. A hypointense signal was seen on the T1 sequence (fig. 3), and a hyperintense signal was seen on the STIR image (fig. 4). A moderate peripheral enhancement was observed after an intravenous injection of paramagnetic contrast material (fig. 5, fig. 6).

Following the results of the imaging studies, surgical excision was recommended as the best treatment of choice. Total parotidectomy with incomplete tumor resection and facial nerve preservation was performed. Subsequently, a biopsy confirmed a ‘fragment of acinic cell carcinoma’ (fig. 7, fig. 8). As a result, the Head and Neck Tumor Board recommended complementary radiation therapy based on the previous surgical decision of incomplete tumor resection. Currently, the patient has remained disease-free with no signs of recurrence.

Discussion

ACCs are a type of uncommon salivary gland tumors that were considered benign. However, in 1953, Buxton demonstrated their ability to metastasize and recur locally [1]. ACCs comprise approximately 7–15% of all malignant tumors arising in major salivary glands. The majority of ACCs (almost 80%) occur in the parotid gland, and approximately 13–17% involve the minor intraoral salivary glands [2, 3]. The female to male ratio is approximately 1.5:1, and the age distribution is fairly even from the second to the seventh decades of life, with a slight peak in the fifth and sixth decades [4, 5]. Possible causes of ACC include previous radiation exposure and familial predisposition [6].

Histopathologically, the tumor is well circumscribed with a distinct capsule, may be solid or cystic and distinct morphological growth patterns are seen. These are described as solid, microcystic, follicular and papillary cystic tumors [7, 8]. They are composed of diverse cell types and include acinic cells, vacuolated cells, intercalated cells, nonspecific glandular cells and clear cells. Large lobules or nests of tumor cells with little intervening stroma are characteristics [9–11]. The histologic grading of acinic cell carcinomas is controversial, and unlike clinical staging, histomorphology has not proven to be reliable in predicting its behavior [12].

No known imaging characteristics of parotid gland ACCs have been found on CT, MRI, or ultrasound imaging. The diagnosis of ACCs using only imaging studies is complex due to its great radiologic similarity with benign tumors [4]. Ultrasonography, which is an easy noninvasive and widely available test, is useful in evaluating tumor size, location, and nature. In addition, it is also used to perform ultrasound-guided fine needle aspiration biopsies. A CT
scan usually demonstrates a slight contrast enhancement and may be appropriate for the evaluation of tumor size, involvement, relationship to facial nerves, other structures and distant metastasis. ACCs usually demonstrate nonspecific signal intensity patterns on MRI, and low T1 and T2 signals can be detected on some images. The signals correlate with the histology, suggesting vascularity, hemosiderin deposition, fibrosis and calcification within the tumor itself [6].

The treatment of choice for ACCs is the complete surgical excision of the tumor by intraoral or extraoral approach. This is determined by the location and the spread of the tumor [2]. Radiation therapy should be considered in cases of poor prognosis, positive surgical margins, stage T3 or T4 tumors, high histologic grade, multiple positive lymph nodal involvement and vascular or perineural invasion [13].

Cure rates have been found to be 76, 63, and 55% at the 5-, 10-, and 15-year marks, respectively. Cervical lymph node metastasis occurred in 3.8–16% of the patients. Distant metastases (liver, lungs and orbit) have been reported between 7 and 29%. The death rate due to ACCs varies from 1.3 to 26%. The recurrence rate for these tumors ranges from 30 to 50%, and when the deep lobe of the parotid gland is reached, local recurrences are higher than in superficial tumors (72 and 18%, respectively) [11, 14, 15].

The 5-year local regional control rate in salivary gland cancer patients managed exclusively by radiation therapy (inoperable tumors, refusal of surgery or incompletely resectable tumors) ranged from 56 to 70% at 5 years, and from 40 to 59% at 10 years. Local regional control rates following surgery and postsurgical radiotherapy ranged from 88 to 91.4% at 5 years, and were 83 and 67% at 10 and 20 years, respectively [16].

Conclusion

ACCs are a type of uncommon salivary gland tumors with the ability to metastasize and recur locally. They often occur between the fifth and sixth decades of life. The diagnosis of ACC using only imaging studies is complex due to its great radiologic similarity to benign tumors. The treatment of choice is complete surgical excision. Radiation therapy should be considered in cases with positive surgical margins, multiple positive lymph nodes and vascular or perineural invasion.

Disclosure Statement

The authors have no conflicts of interest to disclose.

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Fig. 1. CT soft tissue window showing an iso-hypodense nodular mass in the right parapharyngeal space.
Fig. 2. CT soft tissue window after intravenous injection showing moderate enhancement and displacement of the cervical great vessels.
Fig. 3. MRI T1 sequence: hypointense signal of the solid mass.

Fig. 4. MRI STIR sequence: hyperintense signal of the mass is characteristic of a cystic component.
Fig. 5. Gadolinium-enhanced and fat-saturated MRI T1, axial view: moderate peripheral enhancement.

Fig. 6. Gadolinium-enhanced and fat-saturated MRI T1, coronal view: encapsulated mass with mass effect in mucous pharyngeal space.
Fig. 7. HE staining.

Fig. 8. Carcinoembryonic antigen-positive staining.