CASE REPORT

Parapharyngeal Acinic Cell Carcinoma: Initially managed as a Case of Peritonsillar Abscess

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Abstract

We are highlighting an unusual presentation of parapharyngeal acinic cell carcinoma. The patient presented with jaw pain, trismus and a peritonsillar bulge diagnosed as peritonsillar abscess. Medical management was initiated but failed. Magnetic resonance imaging (MRI) was done which revealed abscess formation. Incision and drainage was performed without resolution. Further evaluation lead to the diagnosis and surgical management of parapharyngeal acinic cell carcinoma.

Keywords: Acinic cell carcinoma, Head and neck cancer, Minor salivary gland, Parapharyngeal cancer.

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Introduction

Acinic cell carcinoma is uncommon accounting for 1–3% of head and neck tumors. It arises from differentiation of salivary gland acini and is more commonly found in the parotid gland. Approximately 90% of ACC occurs in the parotid gland with the rest distributed among the submandibular gland and minor salivary glands.[1] Occurrence in the parapharyngeal space (PPS) is even less common. Primary tumors of the PPS are rare accounting for 0.5–1.5% of head and neck tumors. Only 2% of PPS tumors are ACC.[2] Acinic cell carcinoma in the head and neck usually presents as a slow-growing mass with an indolent course.[1,3,4] For tumors in the PPS, the most common presentation is a neck or oropharyngeal mass and trismus is rarely seen and hardly ever documented in PPS tumors.[2] Suspected PPS tumors are best reviewed with imaging such as MRI or CT scans. Magnetic resonance imaging is the preferred imaging modality as it provides better localization and soft tissue extension. Localization can help differentiate possible etiology as prestyloid tumors are more commonly salivary in origin while poststyloid tumors are neurogenic in origin.[2,5] Acinic cell carcinoma usually appears as a single solid cystic expansive mass with peripheral rim enhancement on MRI.[6] Treatment of ACC is surgical resection with a wide margin.[1,4,6] Parapharyngeal space tumors are best approached *via* a cervical-transparotid or transcervical approach.[2,5] Postoperative radiation therapy is not usually planned as ACC is considered a low-grade malignant tumor.[1] Adjuvant radiotherapy is recommended in the presence of positive surgical margins, tumor spillage, multiple lymph nodes, vascular or perineural invasion.[6,7]

Case Description

A 53-year-old male presented in our hospital with a 4-day history of left jaw and ear pain. This was associated with undocumented fever wherein the patient self-medicated with paracetamol. The following day, the patient noted persistence of pain, but was now associated with difficulty opening the mouth. Patient was still able to tolerate regular diet without difficulty of swallowing. Paracetamol was continued which did not afford relief hence subsequent consult in the emergency room. Physical examination revealed trismus, a left peritonsillar bulge and a rightward deviated uvula. No neck swelling or cervical lymphadenopathy was noted. Patient was admitted as a case of peritonsillar abscess and started on intravenous clindamycin, paracetamol, and parecoxib. Complete blood count revealed an elevated white blood cell count. No febrile episodes were recorded during the patient's hospital stay. Resolution of pain was noted after the 2nd hospital day but trismus and the left peritonsillar bulge persisted. Patient was discharged and advised to continue antibiotic treatment until follow-up the week after.

On follow-up, the patient had no recurrence of pain but still had persistent trismus and left peritonsillar bulge. An MRI with contrast was done in an outside diagnostic imaging center which revealed a hypointense mass with peripheral rim enhancement measuring approximately 4.97 × 3.4 × 4.2 cm was signed out as peritonsillar abscess, left (Fig. 1). Patient was immediately admitted for administration of intravenous piperacillin-tazobactam and incision and drainage. Intraoperatively, minimal discharge was expressed and sent for routine gram stain and culture. Soft tissue specimen was also retrieved and sent for routine histopathology. Only minimal increase in mouth opening was noted postoperatively. No organisms were isolated on gram stain and culture. Histopathology revealed predominantly necrotic tissue with dystrophic calcifications.

On subsequent follow-up, persistence of trismus and left peritonsillar bulge was noted. Computed tomography scan of the neck with contrast was requested and compared with the MRI findings. A minimally enhancing hypodense, soft tissue mass centered in the left anterior carotid space with minimal rim calcifications measuring approximately 5.0 × 3.7 × 4.3 cm was noted (Fig. 2). After discussing the case with the radiologist,

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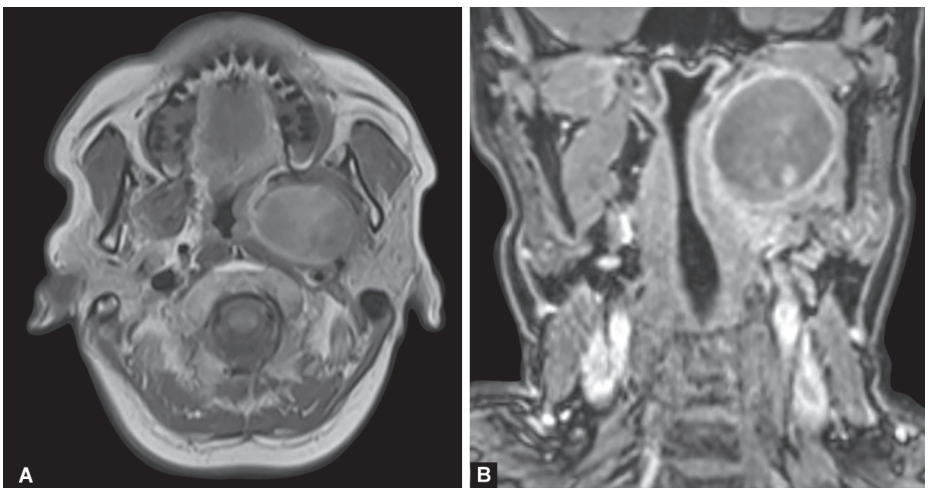
the final impression was a cystic neurogenic tumor. Admission and subsequent excision *via* a transcervical incision were done. Specimen was sent for routine histopathology. Gross examination of the specimen revealed a 5.0 × 3.6 × 2.5 cm pink tan, irregularly ovoid, rubbery tissue with a cystic cavity measuring 4.0 × 2.0 cm filled with yellow-brown pasty material. Extensive necrosis and hemorrhage, approximately 90% of its entirety, were noted. Diagnosis was adenocarcinoma, either ACC or polymorphous low-grade adenocarcinoma. Surgical margins were negative for tumor but lymphovascular invasion was identified.

Slide review of the surgical specimen was performed wherein a diagnosis of ACC was given. Immunohistochemistry staining was done with Discovered on GIST-1 (DOG1) which confirmed the diagnosis of ACC. Patient subsequently underwent 30 fractions of adjuvant radiotherapy receiving 62.5 Gy to the tumor bed and 54 Gy to the ipsilateral neck, levels II–IV, due to the presence of lymphovascular invasion on the final histopathologic report. Latest MRI done 1 year postoperatively was negative for any mass lesion or recurrence. The patient has been on follow-up for 2 years without any new subjective complaints or recurrence.

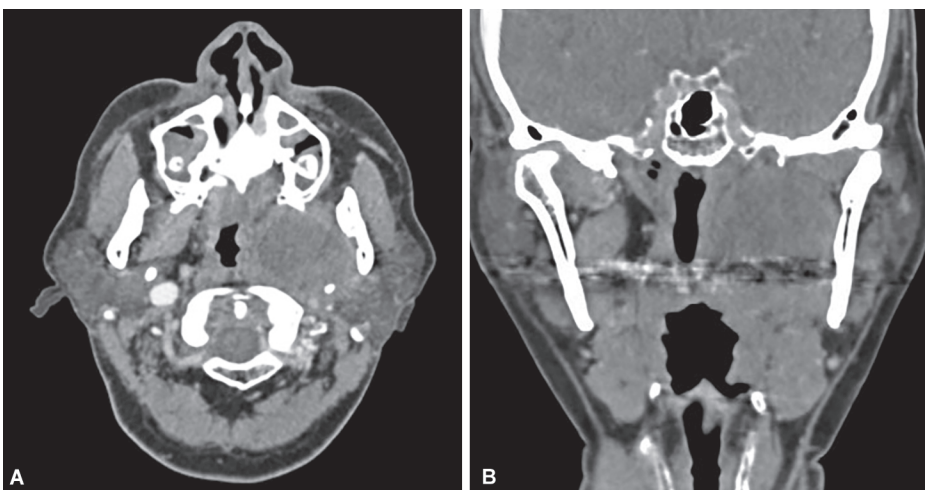
Discussion

Acinic cell carcinoma is a rare epithelial neoplasm of the salivary glands belonging to the family of adenocarcinomas which arises from differentiation from luminar acinar cells.[8,9] Malignant salivary tumors with acinar cell differentiation were first discovered and described by Godwin et al. in the early 1950s when he reported 27 cases of acinic cell adenocarcinomas of the parotid gland.[10] The tumors examined resembled mixed tumors without the myxomatous appearance with occasional cystic and necrotic areas. The lack of histopathologic similarity to other recognized salivary gland tumors, presence of intercellular vacuoles, and a positive periodic acid–Schiff staining has led to the conclusion that these tumors arise from acinar cells.[10] Prior available literature described such tumors as benign adenomas or tumors.[9,10] Due to the documented cases of recurrence, lymph node metastases, and distant metastasis have resulted in consideration of a low-grade malignant process.[9,10]

Clinical presentation of ACC is mostly a painless visual or palpable mass.[2] In our patient, the primary presentation of pain may be secondary to an overlying infection, which resolved



Figs 1A and B: Magnetic resonance imaging of the neck with IV contrast. (A) T1-weighted image axial and (B) T1-weighted coronal image showing a hypointense mass with peripheral rim enhancement measuring approximately 4.97 × 3.4 × 4.2 cm



Figs 2A and B: Computed tomography scan of the neck with IV contrast. (A) Axial image and (B) coronal image showing a minimally enhancing hypodense, soft tissue mass measuring approximately 5.0 × 3.7 × 4.3 cm

after antibiotic treatment. The persistence of a parapharyngeal bulge after the resolution of pain prompted further diagnostic work-up. Identification of a neoplastic process deterred due to nonspecific imaging characteristics of ACC on both CT and MRI.[6,9,11] The utility and significance of MRI for work-up of neoplasms are still considered superior to CT scan due to the invaluable information it provides regarding tumor extent and relationship to the surrounding soft tissue structures.[5] It was just unfortunate that the lesion appeared similar on MRI to a peritonsillar abscess due to the increased signal on fluid-sensitive imaging with peripheral wall enhancement.[12] Nevertheless, radiologic imaging alone would be insufficient in diagnosis of acinic cell tumors as tissue correlation would be necessary.

Fine-needle aspiration has been well established as an effective procedure in the diagnosis of salivary gland tumors.[9] Cytologically, ACC would show presence of acinar differentiated tumor cells. The location of the tumor in our patient prevents proper sampling *via* fine-needle aspiration biopsy. Tissue sampling was done and sent for histopathology but was inconclusive. Final histopathology of the specimen was initially inconclusive with differentials of ACC and low-grade adenocarcinoma. Definitive diagnosis of ACC proves to be a challenging task due to variable microscopic morphologies.[8] Histopathologic similarities are seen with mucoepidermoid carcinoma and low-grade adenocarcinoma of the salivary gland.[9] Immunohistochemical staining has proven to be useful in differentiating salivary gland carcinomas. Two of the identified genes useful for immunohistochemical staining are DOG1 and P63. Discovered on GIST-1 is a calcium-activated chloride channel protein expressed and required for normal salivary secretion which has been positively localized in salivary acini with diminished expression proximally at the level of the intercalated ducts.[8] Both markers could be used to help in arriving at a definite diagnosis. Discovered on GIST-1 was found to be diffusely positive in ACC;[8,13] mostly negative in mucoepidermoid carcinoma;[8] and negative in adenocarcinoma.[13] P63 expression was found to be uniformly positive in mucoepidermoid carcinoma and negative in ACC.[8] Utility of immunohistochemistry staining should be used accordingly based on the diagnostic considerations. For our patient, DOG1 was utilized to confirm ACC as the next consideration was low-grade adenocarcinoma. P63 staining was not requested as mucoepidermoid carcinoma was not part of the diagnostic considerations.

Treatment of choice for ACC remains to be complete surgical excision with negative surgical margins.[1,4,6,9] Adjuvant chemotherapy is not recommended if without distant metastasis[7] and is mainly reserved for pain relief or partial response to treatment.[9] The presence of adverse features such as intermediate to high-grade tumor, T3–T4 tumors, close or positive surgical margins, neural invasion, lymphatic invasion, vascular invasion, and lymph node metastases warrant adjuvant radiotherapy.[6,7,9] Lymphovascular invasion was identified in our patient hence the need for adjuvant radiotherapy for better locoregional control. Despite adequate management, ACC has been documented to have a variable tendency to recur. ACC of the major salivary glands have recurrence rates of 30–50%[6] and may occur up to as late as 30 years from initial presentation[9] whereas those of the minor salivary glands rarely recur.[9,14] Prognosis after adequate treatment of ACC is estimated to be around 91% at 5 years and 88% at 10 years post-treatment.[6,9]

Our patient presented with a rare form of salivary gland carcinoma in a rare location. The clinical presentation alone of ACC is indolent, and given the location of the tumor in the PPS, may come to medical attention at an even later time. The onset of an infection helped identify the presence of a PPS ACC. Incidental discovery of ACC is not unusual and has been documented in various locations.[15,16] A strong clinical suspicion and review of diagnostic imaging is imperative in proper management. Careful review of the CT scan and MRI for resectability was done wherein the decision to perform a diagnostic surgical excision rather than a repeat biopsy was made. Fortunately, despite the lack of a preoperative biopsy, excision with negative margins turned out to be the definite surgical management.

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