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Case Report

Carcinosarcoma of the parotid gland with mucoepidermoid carcinoma component

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ABSTRACT

Carcinosarcoma is a biphasic malignant tumor composed of both carcinomatous and sarcomatous components. First cited in 1951 [1], there have been few cases of this malignant mixed tumor described in the literature. The typical patient presentation is that of an enlarging facial mass in the area of the parotid gland. Systemic symptoms are often absent. Time to initial presentation ranges from months to years. Physical examination findings include swelling and enlargement in the area of the parotid gland, facial nerve deficits, and possible cervical lymphadenopathy. Routine laboratory values (eg, blood counts and electrolytes) usually remain normal, however, nonspecific inflammatory markers (eg, erythrocyte sedimentation rate) may be elevated.

Often times the first step in diagnostic evaluation is computed tomography scan with intravenous contrast. Computed tomography of the head/neck can identify malignant features such as poorly defined margins and calcifications. Magnetic resonance imaging is often performed to better evaluate for soft tissue and perineural invasion. It is important to note that these tumors can be mistaken for abscesses on imaging [2]. Ultimately pathological evaluation with immunohistochemical analysis is required to confirm the diagnosis.

We present a case of a 70-year-old male who initially presented with a painless neck mass. To the best of our knowledge, this is the first case of mucoepidermoid carcinoma associated with carcinosarcoma reported in the parotid gland.

Clinical report

A 70-year-old male presented to the Otolaryngology clinic at Cooper University Health System in February 2019 as a referral from another medical practice in southern New Jersey with a right-sided neck mass. The patient had first noticed a small painless mass in his neck approximately 3 months prior to initial presentation. The mass had slightly increased in size and was occasionally painful. He denied facial symptoms including weakness and paresthesia. He also denied fevers, chills, night sweats, and weight loss. Other than a history of

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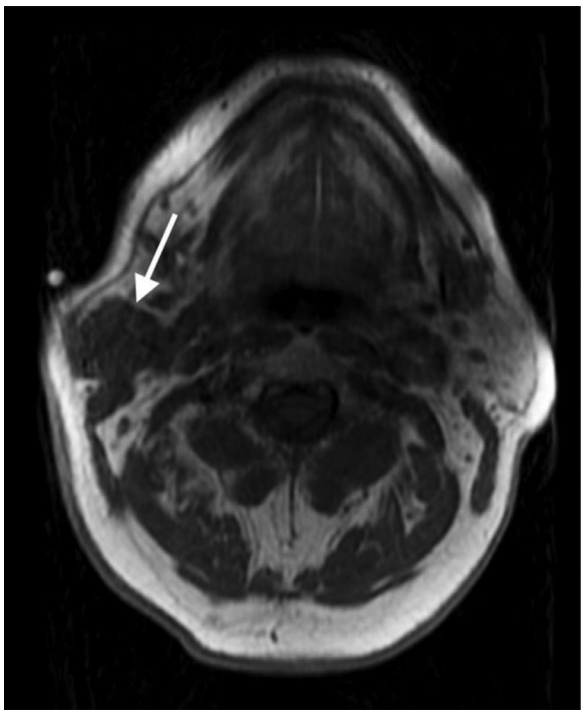

Fig. 1 – T1 axial image of the initial MRI examination which revealed a hypointense mass in the right parotid gland (*arrow*).

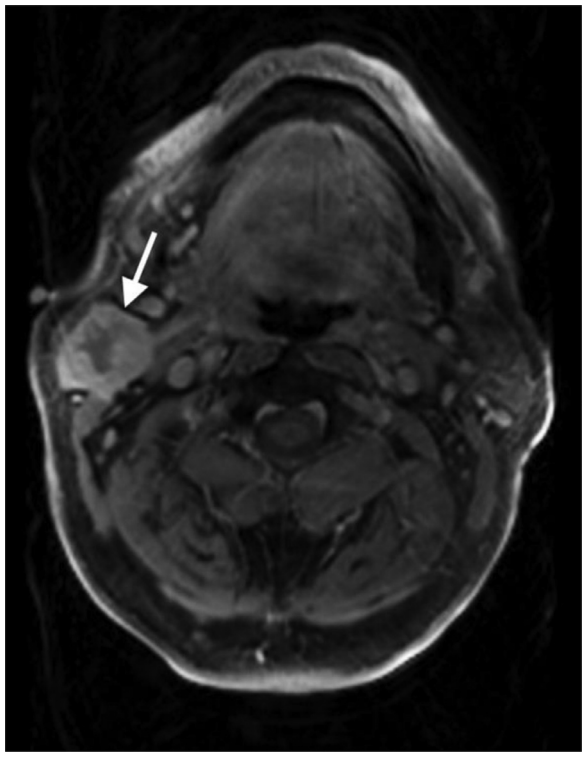

Fig. 2 – Postcontrast T1 axial image of the initial MRI examination. The right parotid mass enhances heterogeneously and demonstrates central necrosis and irregular borders. The contour suggests a malignant process.

hypertension, the patient's past medical history was unremarkable. He was a former 30 pack-year smoker and had a remote history of recreational drug use. Physical examination was notable for a firm, large, well-circumscribed right parotid mass.

There were no overlaying skin changes. There was also no lymphadenopathy of the neck. Cranial nerves II-XII were grossly intact.

An MRI neck with and without contrast performed at the referring medical center revealed a 3.3 × 3.3 × 3.3 cm enhancing tumor in the superficial lobe of the right parotid gland, with irregular margins and possible central necrosis (Figs. 1 and 2). PET-CT scan performed 2 months after this MRI revealed intense hypermetabolic uptake within the right parotid mass with a SUV max of 10.1. The mass measured 4.6 × 4.3 cm in greatest dimension. It extended toward the right parapharyngeal fat plane posterior to the angle of the mandible and displaced the sternocleidomastoid muscle posteriorly. There was no evidence of local, regional or distant metastatic disease (Fig. 3).

Repeat MRI neck without contrast (patient preference) performed 2 months from the initial study revealed an enlarging 4.4 × 4.7 × 5.0 cm well-circumscribed mass with lobulated contour; heterogeneous, hypointense on T1 and heterogeneous, intermediate signal on T2. There was central necrosis, which had progressed from initial MRI. There were no signal abnormalities present in the skull base or any other soft tissue masses in the neck (Fig. 4). Ultrasound-guided fine needle aspiration of the right parotid mass with cytology was per-

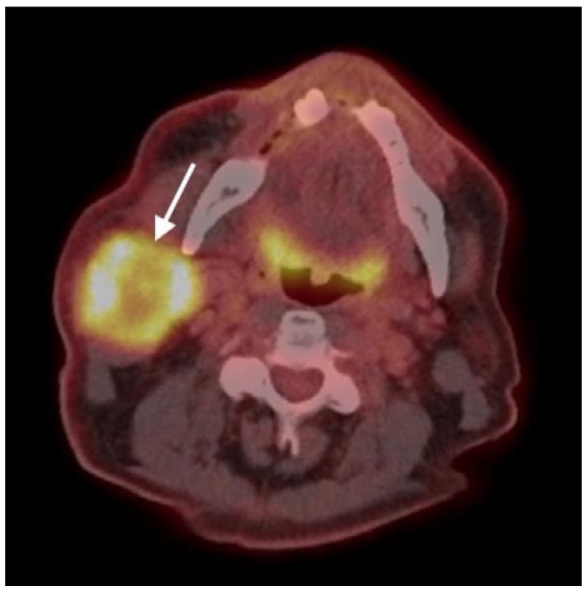

Fig. 3 – PET-CT image which exhibits exquisite hypermetabolism of the right parotid mass with central necrosis. No locoregional or distant metastases were evident on PET-CT.

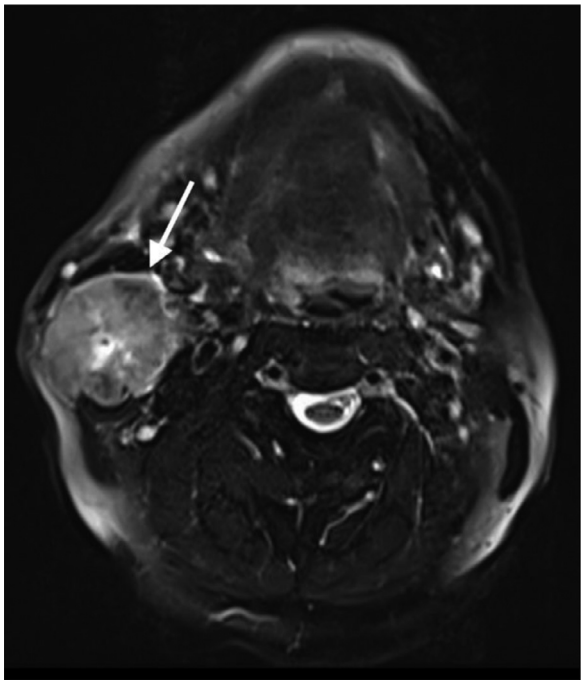

Fig. 4 – T2 axial image of the follow-up MRI study which demonstrates interval enlargement of the mass, progressive irregular contour and central necrosis.

formed and revealed likely high-grade mucoepidermoid carcinoma.

The patient subsequently underwent total right parotidectomy with facial nerve dissection and preservation plus right modified radical neck dissection with sacrifice of the sternomastoid muscle. Grossly, the tumor measured 5 cm. It was firm and pale-tan in appearance with a rim of benign salivary gland tissue at the periphery. No necrosis was noted (Fig. 5). Microscopically, the tumor consisted of 2 intermingled components: (1) moderately to poorly differentiated squamous cell carcinoma forming small clusters to large islands with central necrosis, and (2) high grade sarcomatous spindle cells forming irregular fascicle with buzzard nuclei and abundant abnormal mitoses.

The tumor stroma was dense and fibrotic with focal coarse hyalinization, but no definite osteoid, chondroid, or rhabdoid differentiation was identified (Fig. 6). The tumor focally invaded into benign parotid parenchyma. Immunohistochemically, the carcinoma component was positive for AE1/3, CAM5.2, CK5/6, CK903 and P40, and focal intracellular mucin was also identified by mucicarmine stain (Fig. 7). The sarcomatous component was immunostained for Vimentin, SMA, and SATB2, while no expression was noted for Desmin, Calponin, Myogenin, Pan-melanoma, SOX-10, or S-100 (Fig. 8). The morphology and special study results were consistent with carcinosarcoma with mucoepidermoid carcinoma and undifferentiated sarcoma. No regional lymph node metastasis was found.

Subsequently, the patient received adjuvant external beam radiation therapy which began 6 weeks after surgery. Radio-

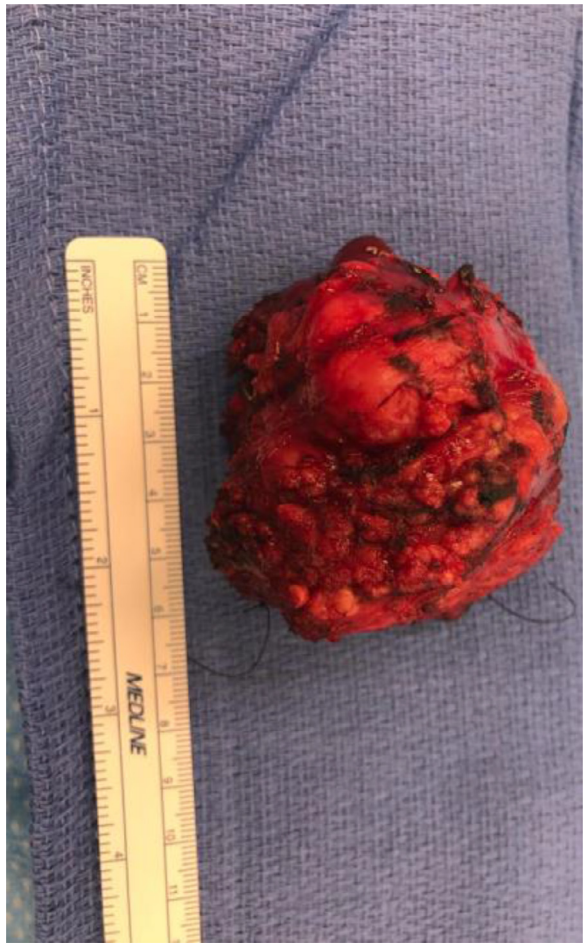

Fig. 5 – Gross specimen of parotid tumor.

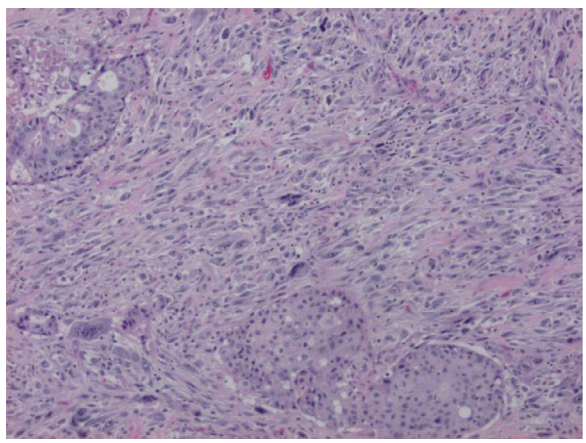

Fig. 6 – Salivary gland carcinosarcoma. The carcinoma component shows moderate squamous differentiation with focal necrosis and cytoplasmic vacuoles, while the highly atypical spindle cells between the carcinomatous islands indicate high-grade sarcoma. HE X100.

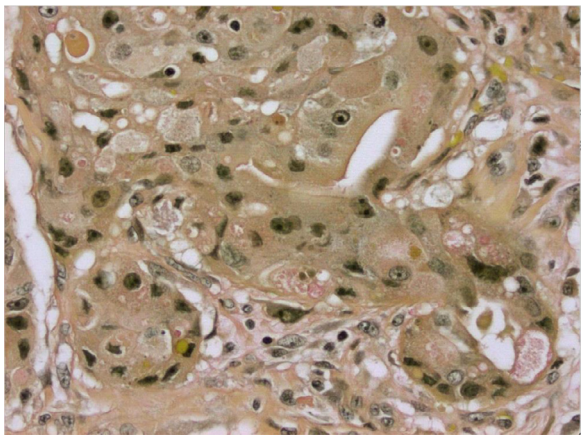

Fig. 7 – Salivary gland carcinosarcoma. Intracellular mucin production is identified in the carcinoma component (pink droplets), supporting mucoepidermoid carcinoma. Mucicarmine stain. X400. (Color version available online.)

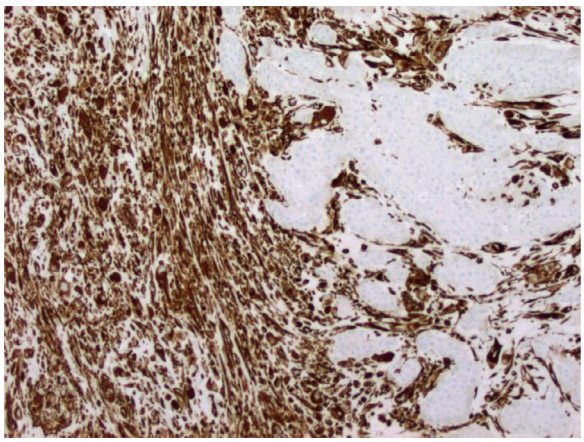

Fig. 8 – Salivary gland carcinosarcoma. The sarcomatous component on the left strongly and diffusely expresses vimentin, a marker for mesenchymal differentiation, while the carcinoma component shows negative immunoreaction. Immunohistochemistry, X100.

therapy was recently completed over the course of 7 weeks and the patient tolerated the treatment well. The patient is scheduled for follow-up PET-CT 3 months after the completion of radiation therapy.

Discussion

Carcinosarcoma is an exceedingly rare tumor of the salivary gland composed of both malignant epithelial and mesenchymal elements [1]. These mixed tumors can arise de novo or from pre-existing pleomorphic adenoma [3]. Its incidence is reported between 0.04% and 0.16% of all salivary gland tumors with the parotid gland being the most affected site [4]. Radiographic imaging is the initial diagnostic test. MRI with contrast is the gold standard and is more accurate than CT scan due to better soft tissue resolution and ability to detect deep lobe involvement as well as perineural invasion. Findings on MRI predictive of malignancy include a well-circumscribed mass with a lobulated contour, heterogeneous, hypointensity on T1, hetereogeneous, intermediate signal on T2, and central necrosis. The presence of either T2 hypointensity or ill-defined margins postcontrast administration has decent sensitivity (70%) and specificity (73%) for predicting malignancy of parotid tumors [5]. It has been reported that parotid carcinosarcomas that parotid carcinosarcoma may be miscontrued as an abscess on imaging studies due to cystic degeneration or necrosis [2].

Patients with carcinosarcoma have a 5-year overall survival of 37% and 5-year disease-specific survival of 62% [6]. Due to such a poor prognosis and high chances of recurrence and metastases, histopathology is warranted to confirm the diagnosis. Histologically, the most common epithelial components include adenocarcinoma, squamous cell carcinoma, and ductal carcinoma, while the most common mesenchymal components include chondrosarcoma, fibrosarcoma, and liposarcoma. To the best of our knowledge, this is the first case of mucoepidermoid carcinoma associated with carcinosarcoma reported in parotid gland, and its impact on tumor behavior is unknown. Marcotullio et al. recently described the first case of carcinosarcoma with mucoepidermoid carcinoma in the submandibular region of salivary glands. Postoperatively, that patient underwent intensity-modulated radiation with 66 Gray in 33 cycles, and no disease recurrence was found at 6-month follow-up [7].

Conclusion

Carcinosarcoma is a clinically aggressive disease with high risk of recurrence. MRI with contrast is the imaging modality of choice, and findings suspicious for malignancy include a well-circumscribed mass with a lobulated contour, heterogeneous, hypointensity on T1, hetereogeneous, intermediate signal on T2, and central necrosis. This is the first reported case of mucoepidermoid carcinoma associated with carcinosarcoma in the parotid gland.

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