

Mammary Analogue Secretory Carcinoma of the Parotid Gland: A Case Report



Casey Fisher DO, Julia Pfaff DO, Matthew Nagorsky MD
Drexel University College of Medicine, Philadelphia College of Osteopathic Medicine



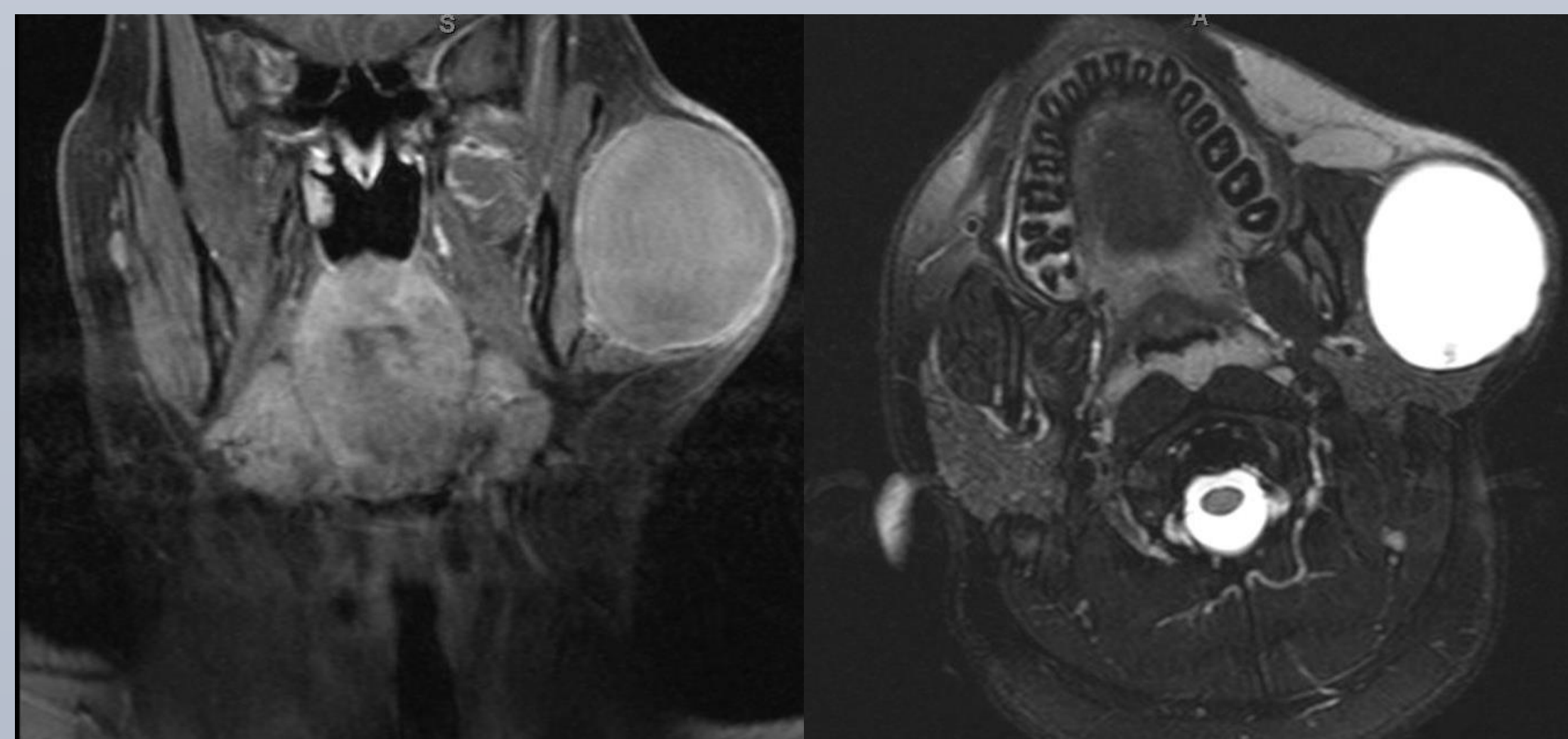
Introduction

Mammary analogue secretory carcinoma (MASC) is an exceedingly rare salivary gland tumor recently described in 2010 by Skalova et al.¹ The tumor relates the morphology and genetics of an equally rare malignancy of the breast, secretory carcinoma. MASC has been classified in the past with salivary tumors of similar morphology. Reported cases of MASC encompass a wide clinical course range, from indolent to aggressive. Due to MASC being a newly reported and rare entity of salivary gland malignancy there is very limited knowledge on clinical course and prognosis. In this case report we present a 22 yo AAM with a slowly growing parotid lesion.

Case History

A 22 yo African American male presented with a 3 year history of a painless, slow-growing left parotid mass. He denied fever, chills, weight loss, dysphagia, or voice changes. Physical exam revealed a 7 cm compressible, fluctuant lesion of the left parotid gland. There was no facial nerve deficit. MRI of the neck demonstrated a 6.5 x 5.8 x 6.5 cm well-circumscribed cystic lesion of the left superficial parotid. No cervical adenopathy was noted. FNA resulted in decompression of the lesion, producing approximately 60cc of serous fluid. Cytology was consistent with a cystic lesion without evidence of malignant cells. The patient was taken to the operating room on 11/27/15 for superficial parotidectomy. Pathologic analysis revealed a well circumscribed cystic lesion measuring 3.5 x 3.4 x 1.7cm with negative margins at 2mm. There was no evidence of extraparenchymal or lymphovascular extension. FISH analysis performed at Johns Hopkins University indicated the presence of ETV6 rearrangement, confirming the diagnosis of mammary analogue secretory carcinoma. The patient was staged T2N0M0. The case was presented at our institution's multidisciplinary head and neck tumor board. The panel concluded that no further surgical intervention or postoperative chemoradiation was indicated at this time. The patient will be followed with clinical surveillance and serial imaging to monitor possible recurrence.

Radiology



MRI Neck. 6.5 x 5.8 x 6.5 well-circumscribed cystic lesion of left superficial parotid gland. Temple Hospital. Phila, PA

Discussion

Mammary analogue secretory carcinoma (MASC) is a recently reported minimally invasive salivary gland tumor with histologic similarities to secretory carcinoma of the breast.² MASC was first reported by Skalova et al in 2010 when they described 16 cases that were previously reported as acinic cell carcinoma (AciCC) and adenocarcinoma not otherwise specified (NOS).^{2,1} Since its initial classification, several small case series have highlighted the immunohistochemical and molecular characteristics of MASC, marking its emergence as a distinct clinical entity.^{3,2,4,5,6,7,8,9,10,11.}

Epidemiology and Clinical Course

In a systematic review of 92 reported cases of MASC, Sethi et al highlighted a slight male predominance (55%) with a mean age at diagnosis of 44.2 years.³ Seventy-one percent of the reported cases were located in the parotid gland, while the minor salivary gland origin was rare.³ The most common clinical presentation is a slow-growing painless lesion, with only one reported case of facial nerve compromise.⁹ Disease progression and survival has varied widely among the existing case reports, ranging from indolent to aggressive behavior.³ In their study of 14 patients, Chiosea et al report a mean disease-free survival of 92 months, while Jung et al report 9 patients with a mean disease-free survival of only 44 months.^{9,11} Only 4 disease-attributed deaths have been reported in cases of advanced distant or locoregional disease.^{1,9}

Treatment

Of the reported cases, surgical excision is the primary treatment modality with highly variable uses of postoperative radiation(RT) or chemoradiation(CRT).³ In the initially reported 16 cases by Skalova et al, 7 patients received postoperative radiation while 9 did not.¹ Of the seven patients who received RT, one died of distant disease, one died after aggressive locoregional recurrence, three remained disease-free after treatment of local recurrence, and two remained free of recurrence or progression. Of the 9 patients who did not receive RT, 7 remained free of recurrence, 1 experienced local recurrence, and 1 was lost to follow up. It is difficult to draw conclusions from their results, however, as radiation dose and delivery were not specified, and the patients were not stratified according to disease staging or tumor characteristics.¹

Histology

Histologically, MASC tumors are comprised of solid, microcystic, and papillary architecture with cells that lack basophilic zymogen granules characteristic of acini.¹ These features help to distinguish MASC from its previous classification as an AciCC or adenocarcinoma NOS. Genetically, the cells demonstrate a t(12;15)(p13;q25) translocation that leads to a *ETV6-NTRK3* fusion product that has not been demonstrated in any other salivary gland malignancy, making its identification an important diagnostic indicator in MASC.¹²

Conclusion

While the histologic and molecular features of MASC have been the emphasis of numerous studies in the pathology literature, its clinical diagnosis and treatment has been underrepresented in the otolaryngology literature. Future studies should emphasize the clinical behavior, risk factors, prognostic indicators, disease stratification, treatment considerations, and long term prognosis of this newly reported salivary gland malignancy.

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