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Case Report

A Rare Case of Myoepithelial Carcinoma of the Maxillary Sinus

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ABSTRACT

Myoepithelial neoplasms are uncommon tumours usually arising within the major and minor salivary glands. They account for only 1% of primary salivary gland tumours, and approximately 10% of these are malignant. We report on a case of this tumour arising in the maxillary sinus, which was managed by staged resection followed by adjuvant chemoradiation.

INTRODUCTION

Myoepithelial cells are found in various salivary gland tumours; however, adenomas composed entirely of myoepithelial cells, called myoepitheliomas, were considered a separate entity in the World Health Organisation's updated histological classification of salivary gland tumours in 1991, with the malignant variant being classified as an intermediate to high-grade tumour.(1) However, myoepithelial carcinoma arising in the maxillary sinus is extremely rare, with only a few cases having been reported worldwide. We present a case of a maxillary myoepithelial carcinoma managed in our facility.

CASE PRESENTATION

A 62-year-old male presented to the Department of Otorhinolaryngology with a four-month history of nasal obstruction, swelling over the left maxilla, anosmia, and loss of vision in the left eye. He also complained of dysphagia and odynophagia. In addition to these symptoms, he reported weight loss of 15 kilograms over the preceding four months. The patient had no significant medical history and was a non-smoker.

On clinical examination, a large mass was noted on the left side of the face, involving the left maxilla and zygomatic arch, with decreased sensation over the mass. The left nasal cavity was obstructed entirely with deviation of the nasal septum. There was proptosis and complete ophthalmoplegia of the left eye with no light perception on assessment of vision. Intraorally, a mucosa-covered mass was present, involving the left buccal mucosa, left alveolar ridge, and the hard and soft palate, with displacement of the uvula to the right. No clinically significant lymph nodes were present.

Imaging was performed, and the computer tomography (CT) scan revealed a large, homogeneously enhancing mass with areas of infarction that completely obliterated the left maxillary sinus, extending to the right and causing erosion of the nasal septum, as well as remodelling of the medial wall of the right maxillary sinus. There was erosion of the orbital floor, with invasion of the orbit and associated proptosis. Superiorly, there was erosion of the floor of the frontal sinus, erosion of the clivus, and obliteration of the sella turcica with intracranial extension into the anterior cranial fossa as well as the middle cranial fossa. The mass was encasing the internal carotid artery and eroded into the left mastoid with erosion of the mandible and subluxation of the temporomandibular joint. Inferiorly, there was an invasion of the soft and hard palate.

The initial biopsy specimen suggested a hyalinising clear cell carcinoma; however, fluorescence in situ hybridisation (FISH) was negative for rearrangement of the EWSR1 gene, and a diagnosis of high-grade myoepithelial carcinoma was made based on the high mitotic rate, nuclear pleomorphism, and presence of tumour necrosis. The specimen was positive for cytokeratin AE1/AE3, smooth muscle actin (SMA), and p16 on immunohistochemistry, but was negative for S-100.

The patient was taken for a staged resection of the tumour in collaboration with the neurosurgical department in our facility. The initial procedure was an endoscopic transnasal debulking of the tumour. This was followed by digital subtraction angiography and particle embolisation of the ascending pharyngeal artery, and the internal maxillary artery was embolised to reduce tumour vascularity.

The patient subsequently had a craniotomy a month after the initial procedure for debulking of the intracranial

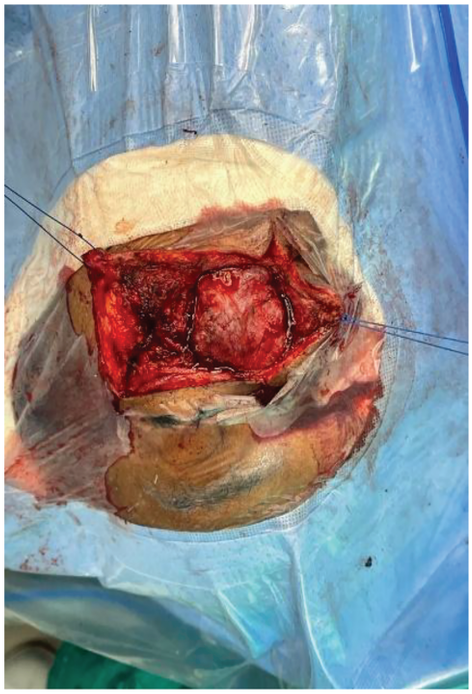

Figure 1: Intraoperative view of the intracranial and peri-orbital tumour component during staged debulking.

Figure 2: Intraoperative view after debulking of the intracranial and peri- orbital tumour component.

component of the mass in the temporal and para-orbital region (Figures 1 and 2). The last stage of the resection was performed three weeks later. It involved a left trans maxillary approach to debulk the tumour, together with closure of the defect, using a titanium mesh mould over the zygomatic, nasal, and maxillary bones. The patient recovered postoperatively in the neurosurgical high-care unit. He was discharged and was scheduled for adjuvant chemotherapy, radiation therapy, and regular follow-up.

DISCUSSION

Myoepithelial carcinoma of the maxillary sinus is a very rare entity, with the first case reported in the English literature occurring in a 67-year-old man in the Netherlands in 1998.(2) Based on our literature review, only 6 cases have thus far been reported. The parotid gland is the most common site for salivary gland myoepithelial carcinoma, followed by the submandibular gland, and a small percentage occurs in the minor salivary glands. Maxillary sinus myoepithelial carcinoma probably arises from accessory salivary gland tissue at this site.(3) The tumour tends to appear in the 40–65 year age group, and there is no clear gender predominance.(2) The presenting symptoms of myoepithelial carcinoma are non-specific and depend on the degree of invasion and destruction of surrounding tissue, and may include facial swelling, nasal congestion, nasal discharge, aural fullness, serous otitis media, hearing loss, and visual disturbances.(3) Imaging helps determine the extent of the tumour and aids in the planning of surgical resection. On CT imaging, myoepithelial carcinoma is isodense to muscle and shows homogenous enhancement, and on magnetic resonance imaging (MRI), it is hypointense on T1.(4)

On immunohistochemical staining, myoepithelial carcinoma is generally positive for pankeratin and vimentin, with variable expression of SMA, calponin, GFAP, and p63. Most reported series describe S-100 positivity in the majority (>90%) of cases. In our case, immunohistochemical analysis demonstrated tumour cell positivity for cytokeratin AE1/AE3 and smooth muscle actin (SMA), with p16 expression, but was negative for S-100 protein. This immunoprofile is therefore somewhat atypical, and the diagnosis rested on the combination of characteristic malignant morphology, including high mitotic activity, nuclear pleomorphism, and tumour necrosis, together with a broader immunohistochemical panel demonstrating epithelial and myoepithelial differentiation.

Surgical resection with negative margins is the treatment of choice for myoepithelial carcinoma;(5) however, in our case, this was not entirely feasible due to the extent of intracranial tumour invasion. Various surgical approaches have been described and used over the years for the resection of sinonasal tumours, and a combined open and endoscopic approach is beneficial in the treatment of some maxillary tumours.(6) This approach was used for our patient in a staged fashion to debulk the tumour, given significant intracranial extension.

The patient underwent adjuvant chemotherapy and radiation therapy. He was reviewed in our outpatient clinic two weeks postoperatively and subsequently at three and six months. At the time of the last documented follow-up, the

patient was clinically stable; however, he has since been lost to follow-up.

CONCLUSION

Maxillary sinus myoepithelial carcinoma is a very rare tumour, and there are currently no established guidelines for treatment. Surgical resection, together with chemotherapy and radiation therapy, is the current mainstay of treatment. Various combinations of these therapies have been used in the few published cases. However, the best treatment method can only be determined with larger numbers over time.

DECLARATION OF CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest that may have inappropriately influenced their writing of this article.

REFERENCES

1. Seifert G, Sobin LH. The world health organization's histological classification of salivary gland tumors: a commentary on the second edition. Cancer. 1992; 70(2):379–385. doi:10.1002/1097-0142(19920715)70:2<379::aid-cncr2820700202>3.0.co;2-c
2. van der Graadt Roggen JF, Baatenburg de Jong RJ, Verschuur HP, et al. Myoepithelial carcinoma (malignant myoepithelioma): first report of an occurrence in the maxillary sinus. Histopathology. 1998; 32(3):239–241. doi:10.1046/j.1365-2559.1998.00382.x
3. Zhou S, Ruan L, Gong L, Wang S. Primary malignant myoepithelioma of the left maxillary sinus: a case report. J Int Med Res. 2008; 36(2):362–365. doi:10.1177/147323000803600221
4. Patil B, Hegde S, Yadav KD. High-grade myoepithelial carcinoma of the maxillary sinus. *Int J Dent Res*. 2017; 5(2):143–144. Available from: https://www.sciencepubco.com/index.php/IJDR/article/view/8039
5. Ghosh A, Saha S, Saha VP, Sadhu A, Chattopadhyay S. Infratemporal fossa myoepithelial carcinoma—a rare case report. Oral Maxillofac Surg. 2009; 13(1):59–62. doi:10.1007/s10006-009-0149-5
6. Chatelet F, Simon F, Bedarida V, et al. Surgical management of sinonasal cancers: a comprehensive review. Cancers. 2021; 13(16):3995. doi:10.3390/cancers13163995