

## Myoepithelial Carcinoma in Maxilla: A Rare Case Report

Chiranjib Das, Mukesh Kumar Singh, Aniruddha Majumder, Soumik Das, Tapan Kanti Hazra

### ABSTRACT

Myoepithelial carcinoma (MC) is an extremely rare neoplasm of salivary gland. It is usually found in parotid gland. Here, we report a rare case of myoepithelial carcinoma of maxillary sinus along with discussion of clinical, histopathological and immunohistochemical characteristics of this rare disease.

**Keywords:** Myoepithelial carcinoma, Maxilla, Histopathological examination, Immunohistochemistry.

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### INTRODUCTION

Myoepithelial carcinoma is defined by World Health Organization (WHO) in 1991 as a rare low-grade malignant neoplasm of salivary gland which accounts for less than 1% of salivary gland tumors. Though the name was coined by Donath et al in 1972, it was likely recognized as early as 1956 and reported under variety of names, such as adenomyoepithelioma, clear cell adenoma, tubular solid adenoma and clear cell carcinoma.<sup>1</sup> The tumor most commonly arises from parotid gland. Very few cases have been reported in literature in palate, gum, sinonasal tract, larynx, lateral wall of nasopharynx and tongue base.<sup>2</sup> Here we present a rare case of myoepithelial carcinoma of maxillary sinus which was diagnosed postoperatively by histopathological and immunohistochemical study.

### CASE REPORT

A 55-year-old male presented to our ENT OPD with gradually progressive nasal obstruction on left side for 6 years. For last 6 months he noticed a gradually progressive swelling in his left cheek. On examination we found a soft, reddish, nonfriable, nontender mass occluding left nasal cavity which appears to arise from lateral wall of left nasal cavity. Nasal septum was deviated to right side. A soft, cystic, fluctuant swelling of 1.5 × 3 cm on left malar prominence with erythema of overlying skin was noted (Fig. 1). We aspirated about 10 ml of pus mixed with fresh blood and send it for culture and sensitivity examination. Thereafter on palpation we appreciated an underlying bony defect on anterior wall of left maxilla 1 cm below infra-orbital margin and 1.5 cm medial to zygomaticomaxillary complex. CECT of paranasal sinuses showed heterogeneously enhancing soft tissue mass involving left maxillary sinus



Fig. 1: Swelling in left malar prominence

invading cheek through eroded anterior wall, left infratemporal fossa through posterolateral wall, ipsilateral nasal cavity through its widened ostium and orbit through eroded roof of sinus. The left nasal cavity is completely filled up with soft tissue mass extending anteriorly into nasal vestibule and posteriorly through choana into nasopharynx up to tubal elevation and superiorly into ipsilateral ethmoidal labyrinth and sphenoidal sinus. Nasal septum is slightly deviated toward right side (Figs 2 and 3). Biopsy was taken from the mass in left nasal cavity endoscopically and histopathological examination showed inverted papilloma with dysplastic changes. The tumor was excised under general anesthesia by Weber-Ferguson incision, removing the whole mass with eroded anterior wall, posterolateral wall and roof of left maxillary sinus keeping intact the lateral part of zygomaticomaxillary complex and suspensory ligaments of left eyeball. The patient's postoperative course was uneventful. Histologically, the tumor was composed of atypical large hyperchromatic polygonal and spindle cells arranged in cords separated by hyaline stroma. There are foci with high mitotic count (Fig. 4). The neoplastic cells express cytokeratin, EMA P63 and calponin and are immunonegative for CD34. It was consistent with the diagnosis of myoepithelial carcinoma. Postoperatively 60 Gy of radiotherapy was given in 30 sessions as advised by oncologist. There was no clinical and radiological evidence of recurrence of tumor in 6 months follow-up.

### DISCUSSION

Myoepithelial carcinoma is a rare malignant neoplasm of salivary gland. It constitutes less than 1% of all salivary

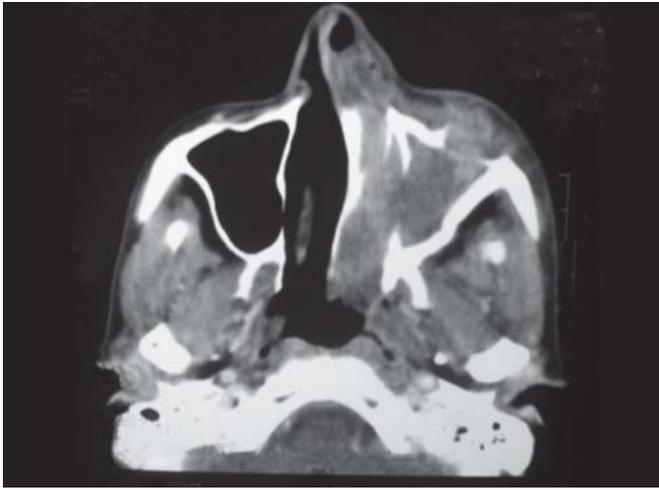


Fig. 2: CECT of PNS (axial view)



Fig. 3: CECT of PNS (coronal view)

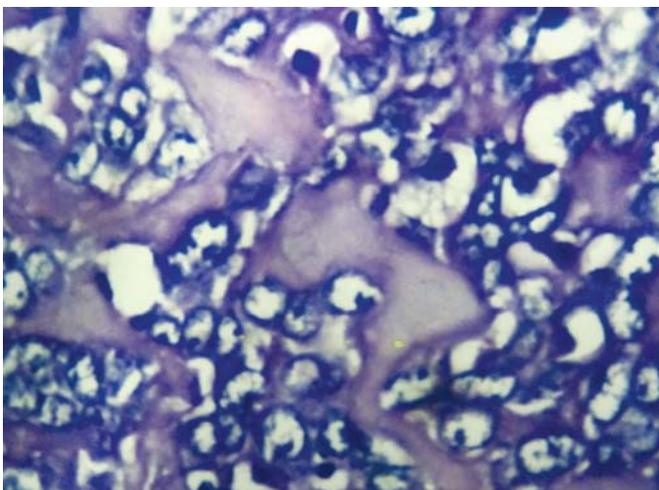


Fig. 4: Eosinophilic cuboidal duct alike cells form lumens and are surrounded by polygonal, clear myoepithelial cells

gland tumors. It is usually found in major salivary gland and parotid gland. However, very rarely it may arise from minor salivary gland located in palate, gum, larynx, lateral wall of nasopharynx, sinonasal tract and tongue base.<sup>2</sup> Women are more frequently affected (2:1). The incidence

peaks between 50 and 60 years of age. Patients typically present with painless slow growing mass, occasionally there is rapid growth, pain or facial weakness.<sup>3</sup> In our case the patient was a 55-year-old male who presented with a slow growing mass in left maxilla which recently eroded the bony walls and extended into surrounding soft tissues. The imaging appearance of myoepithelial carcinoma is not specific.<sup>4</sup> Macroscopically, it is well-circumscribed grayish white to tan white mass.<sup>4</sup> Microscopically, it consists of two cell types. The outer layer of myoepithelial cells surrounds an inner layer of ductal epithelial cells. Myoepithelial cells are large, with clear staining cytoplasm. Epithelial cells are cuboidal with eosinophilic cytoplasm and uniform round nuclei. Cytoplasmic atypia is mild or absent, but occasionally cellular and nuclear pleomorphism and high mitotic figures are present. The duct or gland like lumen may contain eosinophilic proteinaceous material which is reactive to periodic acid schiff staining. Myoepithelial immunohistochemical markers, i.e. smooth muscle cell actin, calponin and p63 highlight the clear myoepithelial cells and ductal epithelial cells react with cytokeratin.<sup>3</sup> If the tumor is accessible for resection, radical resection is the treatment of choice for myoepithelial carcinoma.<sup>4</sup> Though in our case total maxillectomy was not done as preoperative biopsy report was inverted papilloma and during dissection we did not find any involvement of palate, alveolar margin and zygomaticomaxillary complex. Radiation therapy can be used, either alone or in conjunction with and after surgery, particularly for unresectable lesions or metastatic disease. In our case radiation was given as we did not performed radical surgery and the diagnosis of malignant tumor was done postoperatively. Overall prognosis is good with 5 years survival of 80% following complete surgical excision. Recurrences have been reported in as many as 40% of cases, usually within 5 years of initial surgery. Metastasis occurs in about 15% of cases, usually in cervical lymph node, lung, liver and kidney. Fewer than 10% of patients die from disease complication. Large tumor size and rapid growth are associated with worse prognosis.<sup>3</sup>

## CONCLUSION

Though myoepithelial carcinoma is a rare tumor of salivary glands, its occurrence in other sites is still possible. So, it should be kept as a differential diagnosis of any locally aggressive tumor of head and neck region. Careful evaluation and various diagnostic tests are required to confirm the diagnosis as often a preoperative diagnosis is not possible from the clinicoradiological examination. Postoperative histopathological examination and immunohistochemistry are of great value. Most appropriate treatment of this type of tumor is radical resection, if it is

possible and radiation therapy is an alternative where total resection is not possible.

## REFERENCES

1. Alves Muniz Pollianna, Godoy Pina Gustavo, et al. Unusual epithelial myoepithelial carcinoma in palate- case report and immunohistochemical study. *Oral Medicine and Pathology. J Clin Exp Dent* 2010;2(1):22-25.
2. Tuncel Unit, Ergul Gulusan, et al. Myoepithelial carcinoma in nasopharynx: An unusual localization. *Yonsei Medical Journal* 2004;45(1):161-65.
3. Folk S Gretchen, Thompson DR Lester. Epithelial myoepithelial carcinoma. *ENT Journal Pathology Clinic*.
4. Pradhan A Sultan, Desai Meena, et al. Sinonasal epithelial myoepithelial carcinoma: A rare entity. *Indian J Otolarygol. Head Neck Surg* April-June 2007;59:168-70.

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