

Myoepithelial carcinoma of the nasopharynx: Case report of a rare entity

Department of Oral and Maxillofacial Surgery, Sri Guru Ram Das Institute of Dental Sciences and Research, Amritsar, ¹Kidwai Memorial Institute of Oncology, Bangalore, Karnataka, India

Amit Dhawan, Ashok Shenoy¹, D. Sriprakash¹

Address for correspondence:
Dr. Amit Dhawan,
Department of Oral and Maxillofacial Surgery, Sri Guru Ram Das Institute of Dental Sciences and Research, Amritsar, India.
E-mail: surg.amit@gmail.com

ABSTRACT

Myoepithelial carcinomas are rare tumors of salivary glands. Most occur in the parotid gland but a few other sites of origin have also been described. Myoepithelial carcinoma of the nasopharynx has only been reported very few times. Because the lesion is so rare in the nasopharynx, there are no specific guidelines for its treatment. We present a rare case of myoepithelial carcinoma in the nasopharynx and discuss its diagnostic and therapeutic aspects.

Key words: Myoepithelial carcinoma, nasopharynx, surgical management

INTRODUCTION

Myoepitheliomas are relatively rare tumors of the salivary glands comprising only 1% of all. Most of these are benign but malignant variants, called myoepithelial carcinomas (MCs), are even rarer comprising about 10% of myoepitheliomas.^[1,2] Most of these occur in the parotid gland; a few other sites of origin have been described such as lungs, trachea, oral cavity, larynx and breast.^[3,4] To our knowledge there are very few countable reports of MCs of the nasopharynx in the literature which makes the nasopharynx an unusual location for MCs and considered difficult to access for the head and neck surgeon. So, there are no specific indications and guidelines for its management. We present a case of myoepithelial carcinoma in the nasopharynx and discuss its diagnostic and therapeutic aspects.

CASE REPORT

A 60-year-old female reported to the department with a non-painful left laterocervical swelling measuring

3 cm in diameter. The patient complained of nasal blockage, hyponasal speech, conductive hearing loss and ear fullness since two months though the cranial nerve examination results were within normal limits. Posterior rhinoscopy examination revealed a fungating tumor on the roof and left lateral aspect of the nasopharynx. Endoscopic nasopharyngeal biopsy was taken which reported the lesion as a mucus-secreting minor salivary gland tumor. Fine needle biopsy of the nasopharyngeal swelling was performed, and the lesion was diagnosed as myoepithelial cell carcinoma.

Computed tomography (CT) scan depicted an irregular hypodense lesion measuring 4.5/3.8 cm in the left half of the nasopharynx, obliterating the Eustachian tube opening and Rosenmuller fossa. On the left side the lesion was extending to the pterygopalatine fossa and involving the parapharyngeal space. On the ipsilateral side, the submandibular group of lymph nodes was enlarged. There was no intracranial extension noted [Figure 1]. Chest X-ray and Ultrasound (USG) abdomen were performed as a part of the metastatic workup.

The patient underwent excision of the tumor with paramedian mandibulotomy in conjunction with transpalatal approach [Figure 2]. Supraomohyoid neck dissection was performed on the ipsilateral side with classical transcervical incision. Histopathology reports revealed myoepithelial carcinoma of the nasopharynx with infiltrating margins and the presence of numerous sheets and cords of both spindle-shaped and epithelioid neoplastic cells [Figure 3]. Tumor was immunoreactive

Access this article online	
Quick Response Code: 	Website: www.njms.in
	DOI: 10.4103/0975-5950.94484

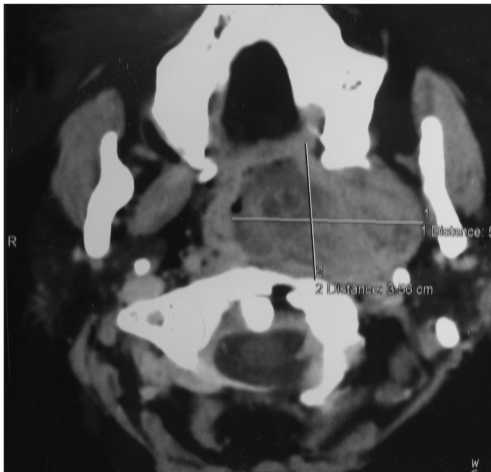


Figure 1: Axial CT scan showing the extent of the tumor in the nasopharynx

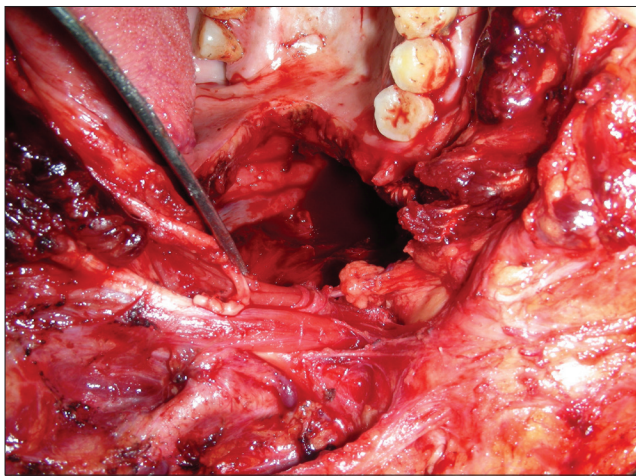


Figure 2: Intraoperative photograph showing the surgical site after excision of the tumor

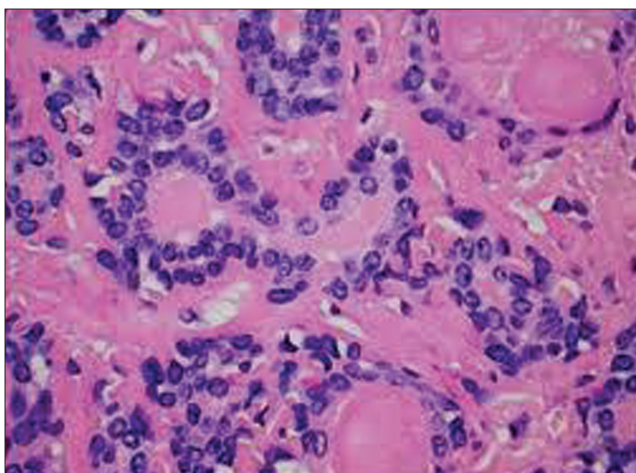


Figure 3: Histopathological picture of myoepithelial carcinoma showing spindle-shaped cells in the background

for actin, smooth muscle actin (SMA), S-100 and cytokeratin (CK) markers. The postoperative period was uneventful and the patient received external beam

radiotherapy to the primary site {70 gy/ 35 fractions} as well as to the neck {56 gy/ 28 fractions} which was started within three weeks of surgery. The patient is on regular follow-up since 28 months with no evidence of recurrence at the loco-regional level till date.

DISCUSSION

MCs are tumors of epithelial origin that may occur with preexisting benign lesions like pleomorphic adenomas or benign myoepitheliomas, but they also may arise *de novo*.^[5] Nilles and associates^[6] and Tuncel and colleagues^[7] reported only two cases of malignant myoepithelioma in the rhinopharynx that were confirmed histologically and immunohistochemically. In 1983, Nofal^[8] described a poorly differentiated spindle-cell carcinoma that could have been the third case of rhinopharyngeal myoepithelial carcinoma.

MC is usually insidious in onset, a painless mass that originates in the parotid gland. However, other sites such as the palate, larynx, gums, retromolar area, and breast have also been reported.^[3,4] In the parotid gland, MCs usually affect patients over 50 years of age with no sex predilection. Because they are relatively painless tumors, their diagnosis can be delayed by months or even years.^[1]

When MCs occur in the nasopharynx, the symptoms are the same as those of other tumors that affect this region, such as nasal obstruction, ear fullness, serous otitis media, and conductive hearing loss. To ensure a correct early diagnosis of nasopharyngeal tumors, the lesion should first be evaluated via fiberoptic nasopharyngoscopy to identify the mass, which is usually covered with unaltered mucosa. Diagnostic imaging (CT and magnetic resonance imaging (MRI)) allows the site and the extension of the tumor to be established, thus permitting a correct surgical approach. The diagnosis can be further ascertained by taking a biopsy specimen for histological confirmation of the presence of a tumor and its type. Histologically, MCs appear to have pleomorphic spindle-shaped or more rounded cells, occasionally with eosinophilic cytoplasm (otherwise known as plasmacytoid cells).^[3] Immunohistochemistry can help to identify myoepithelial differentiation of the neoplastic cells. Chow *et al.*,^[9] and Torlakovic and associates^[10] demonstrated that tumoral cells are immunoreactive to cytokeratin, smooth muscle actin, and S-100 protein. Our patient was no exception, and this profile allowed the final histological diagnosis of myoepithelial carcinoma. The criteria that indicate the malignancy of myoepithelial neoplasms include their destructive infiltrating growth, cellular pleomorphism, necrosis, and an increase in mitotic activity.^[11] Locally, this type of tumor is particularly destructive, but

its clinical and biological features are not yet fully understood. Some authors believe that metastases are uncommon,^[1] while others have reported metastases in 30% of the cases.^[9] It has been reported that MCs in the parotid may spread to the submandibular and supraclavicular lymph nodes, hence requiring neck dissection procedures.^[4]

The treatment of choice for MCs is surgical excision, but there are no specific guidelines for the treatment of nasopharyngeal MCs. However, it seems reasonable to apply the criteria used for MCs involving the most common site, the parotid. So any type of selective neck dissection will prove to be a diagnostic as well as a therapeutic procedure. Unlike certain nasopharyngeal tumors for which chemotherapy and radiotherapy are the preferred treatments in some cases, the application of these treatments to MCs has not been encouraging. The mainstay of success is primary surgical management.^[11] Several surgical approaches have been proposed, depending on the size and site of the tumor.^[6-8,12] We adopted excision of the tumor with paramedian mandibulotomy in conjunction with a transpalatal approach. To our knowledge, this patient is the only case of nasopharyngeal MC treated with this technique. It enabled complete resection of the lesion with negative margins, and there have been no signs of recurrence and metastasis during the 28-month follow-up period with stable dental occlusion, no velo-pharyngeal incompetence and no signs of trismus.

ACKNOWLEDGMENT

We acknowledge Dr P Chavan, Assistant Professor, Department of Head and Neck Oncology and Reconstructive Surgery, for his valuable contribution to the case report.

REFERENCES

1. Sciubba JJ, Brannon RB. Myoepithelioma of salivary glands: Report of 23 cases. *Cancer* 1982;49:562-72.
2. Barnes L, Appel BN, Perez H, El-Attar AM. Myoepitheliomas of the head and neck: Case report and review. *J Surg Oncol* 1985;28:21-8.
3. Dean A, Sierra R, Alamillos FJ, Lopez-Beltran A, Morillo A, Arévalo R, *et al.* Malignant myoepithelioma of the salivary glands: Clinicopathological and immunohistochemical features. *Br J Oral Maxillofac Surg* 1999;37:64-6.
4. Klumpp TR, Mohr RM, Silverman CL, Tang CK, Elfenbein IB, Dardick I. Malignant myoepithelioma of the parotid gland: case report and review of the literature. *J Laryngol Otol* 1995;109:995-8.
5. Young J, Barker M, Robertson T, Nasioulas S, Tannenber A, Buttenshaw RL, *et al.* A case of myoepithelial carcinoma displaying biallelic inactivation of the tumour suppressor gene APC in a patient with familial adenomatous polyposis. *J Clin Pathol* 2002;55:230-2.
6. Nilles R, Lenartz T, Kaiserling E. Myoepithelial carcinoma of the nasopharynx: Case report and review of the literature. *HNO* 1993;41:396-400.
7. Tuncel U, Ergul G, Ozlugedik S, Unal A. Myoepithelial carcinoma in the nasopharynx: An unusual localization. *Yonsei Med J* 2004;45:161-5.
8. Nofal F. Spindle cell carcinoma of the nasopharynx. *J Laryngol Otol* 1983;97:1057-63.
9. Chow LT, Chow WH, Lee JC. Monomorphic epithelioid variant of malignant myoepithelioma of the parotid gland: Cytologic features in fine needle aspiration (FNA). *Cytopathology* 1996;7:279-87.
10. Torlakovic E, Ames ED, Manivel JC, Stanley MW. Benign and malignant neoplasm of myoepithelial cells: Cytologic findings. *Diagn Cytopathol* 1993;9:655-60.
11. Savera AT, Sloman A, Huvos AG, Klimstra DS. Myoepithelial carcinoma of the salivary glands: A clinicopathologic study of 25 patients. *Am J Surg Pathol* 2000;24:761-74.
12. Fisch U. The infratemporal approach for nasopharyngeal tumors. *Laryngoscope* 1983;93:36-44.

How to cite this article: Dhawan A, Shenoy A, Sriprakash D. Myoepithelial carcinoma of the nasopharynx: Case report of a rare entity. *Natl J Maxillofac Surg* 2011;2:207-9.

Source of Support: Nil. **Conflict of Interest:** None declared.