CASE REPORT

Malignant Myoepithelioma of the Rhinopharynx: Case Report

Giuseppe Magliulo, M.D.,¹ Gianna Pulice, M.D.,¹ Massimo Fusconi, M.D.,¹ and Giuseppe Cuiuli, M.D.¹

ABSTRACT

Malignant myoepitheliomas are rare tumors of salivary glands. Most occur in the parotid gland; few other sites of origin are described. Malignant myoepithelioma of the rhinopharynx has only been reported twice. Because the lesion is so rare, there are no specific indications for its treatment. We present a third case of malignant myoepithelioma in the rhinopharynx and discuss its diagnostic and therapeutic aspects.

KEYWORDS: Malignant myoepithelioma, rhinopharynx, surgical treatment

Myoepitheliomas are rare tumors that account for about 1% of all salivary gland tumors.^{1,2} Most are benign, but some have been malignant. The latter, even rarer, entity has been termed malignant myoepithelioma (MM) or myoepithelial carcinoma and represents about 10% of myoepitheliomas. In 1943 Sheldon³ first used the term myoepithelioma and in 1975 Stromeier et al⁴ reported the first case of MM. Since then most MMs described in the literature have been located in the parotid gland. Occasionally, other sites such as the oral cavity, palate, retromolar area, cheek, or larynx have been involved.⁵ MMs are particularly unusual in the rhinopharynx; only two other cases affecting this area have been reported. We present a third case of MM in the rhinopharynx and discuss its diagnostic and therapeutic aspects.

CASE REPORT

In January 1998, a 55-year-old woman sought treatment for a right laterocervical swelling that had appeared in 1991. Its volume had increased progressively to 2 cm in diameter. The patient suffered from nasal obstruction, ear fullness, and conductive hearing loss. The swelling was wooden hard, mobile, and covered with intact skin. It was neither painful nor tender.

Fiberoptic rhinopharyngoscopy revealed a growth in the upper right side of the rhinopharynx descending to the right tonsil, covered with normal mucosa. A biopsy was performed, and the histological examination showed fragments of predominantly mucus-secreting salivary glands substituted with adipose tissue and ectasis of some ducts. Fine

Skull Base, volume 15, number 2, 2005. Address for correspondence and reprint requests: Giuseppe Magliulo, M.D., Via Gregorio VII n. 80, 00165 Rome, Italy. E-mail: giuseppemagliuloorl@yahoo.com. ¹Department of Otorhinolaryngology, Audiology, and Phoniatrics, "Giorgio Ferreri," "La Sapienza," University of Rome, Rome, Italy. Copyright © 2005 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662. 1531-5010,p;2005,15,02,113,118,ftx,en;sbs00443x.

needle biopsy of the laterocervical swelling was performed, and the lesion was diagnosed as myoepithelial cell carcinoma. Magnetic resonance imaging (MRI) showed the presence of pathological tissue with marked dishomogeneity occupying the infratemporal and pterygoid fossae, rhinopharynx, and right parapharyngeal space (Fig. 1).

The patient underwent an infratemporal fossa approach type C for resection of the neoplasm and its involvement with the vertical and horizontal segments of the internal carotid artery. The spinal nerve was preserved through a functional dissection of the neck on the right. The final histological diagnosis noted the presence of numerous sheets and cords of both spindle-shaped and epithelioid neoplastic cells with pale cytoplasm and positive immunohistochemical staining with actin, cytokeratin, and S-100 antibodies. The findings were classified as a pT4pN0pNx myoepithelial carcinoma (Figs. 2 and 3). As of 2004, there have been no signs of recurrence.

Figure 1 Magnetic resonance image of head and neck showing pathological tissue involved with the right rhinopharynx and the parapharyngeal space.

Figure 2 Hematoxylin-eosin staining (original magnification $\times 10$) shows a tumor with clear cells forming lumps separated by fibrous layers.

DISCUSSION

The classification of myoepitheliomas has been under discussion for some years. Some authors consider it a variation of the pleomorphic adenoma² or a form of monomorphic adenoma.⁶ Others define it as a tumor composed solely of myoepithelial cells⁷ or with a less than 10% tubular component.⁶ At

Figure 3 S-100 staining appears to be positive in most cells.

Table 1 Summary of Cases Reported in the Literature

Reference	Tumor	Primary Site	Therapy	Follow-Up	Recurrence
Nofal ¹³	Spindle cell carcinoma	Rhinopharynx	Transpalatal approach	ND	ND
Nilles et al ¹¹	Myoepithelial carcinoma	Rhinopharynx	Paralateral nasal rhinotomy	6 months	No
Tuncel et al ¹²	Myoepithelial carcinoma	Nasopharynx	Midfacial degloving, Le Fort 1 osteotomy, radiotherapy	15 months	Yes
Present case	Myoepithelial carcinoma	Rhinopharynx	Infratemporal fossa approach type C	5 years	No

ND, not determined.

present, myoepitheliomas and MMs are considered two separate clinical pathological entities.⁸ MMs are tumors of epithelial origin that may occur with pre-existing benign lesions like pleomorphic adenomas or benign myoepitheliomas, but they also may arise *de novo*.⁹

Nilles and associates¹¹ and Tuncel and colleagues¹² reported the only two cases of MM in the rhinopharynx that have been confirmed histologically and immunohistochemically. In 1983, Nofal¹³ described a poorly specified spindle-cell carcinoma that could have been the third case of rhinopharyngeal MM (see Table 1).

MMs are usually a slow-growing painless mass that originates in the parotid gland. However, other sites such as the palate, larynx, gums, retro-molar area, and breast have also been reported.¹⁰ In the parotid gland, MMs usually affect patients over 50 years old with no gender preference. Because they are seldom associated with pain, their diagnosis can be delayed by months or even years.²

When they involve the pharynx, MMs provoke aspecific symptoms such as dysphagia, the sensation of a foreign body, or respiratory problems. Retromandibular swelling or medial dislocation of the lateral wall of the pharynx or tonsil has been found on physical examination.

When MMs occur in the rhinopharynx, the symptoms are the same of those of other tumors that affect this region, such as nasal obstruction, ear fullness, serous otitis media, and conductive hearing loss.¹⁴

In order to ensure a correct early diagnosis of rhinopharyngeal tumors, the neoplasm should first be studied closely via fiberoptic rhinopharyngoscopy to identify the mass, which is usually covered with

unaltered mucosa. Any possible adenopathies must be investigated. The next step is imaging (i.e., CT and MRI). CT allows the site and the extension of the tumor to be established, thus permitting a correct surgical approach. MRI is more precise in defining the tumor's relationship with the adjacent structures and internal carotid artery.¹⁴ The diagnosis can be further ascertained by taking a biopsy specimen for histological confirmation of the presence of a tumor and its type.

Histologically, MMs appear to have pleomorphic spindle-shaped or more rounded cells, occasionally with eosinophilic cytoplasm (otherwise known as plasmacytoid cells).¹⁰ Immunohistochemical examination can help to identify myoepithelial differentiation of the neoplastic cells. Chow et al¹⁵ and Torlakovic and associates¹⁶ 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 malignant myoepithelioma.

The criteria that indicate the malignancy of myoepithelial neoplasms include their destructive infiltrating growth, cellular pleomorphism, necrosis, and an increase in mitotic activity.⁸ El-Naggar and colleagues stated that aneuploidy and a high cell fraction in the S phase are signs that predict a tumor's future aggressive behavior.¹⁷ 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,^{2,8} while others have reported metastases in 30% of cases.¹⁵ MMs in the parotid may spread to the supra-clavicular or submandibular lymph nodes, the mastoid or temporal bones, lungs,

or liver.⁵ Ibrahim and colleagues described a case of MM that spread to the liver, pleura, peritoneum, and lumbar spine.¹⁸

The new treatment of choice for MM is surgical excision, but there are no specific guidelines for the treatment of rhinopharyngeal MM. However, it seems reasonable to apply the criteria used for MMs involving other sites. Unlike certain rhinopharyngeal tumors for which chemotherapy and radiotherapy are the preferred treatments in some cases, the application of these treatments to MM has not been encouraging.¹⁹ Several surgical approaches have been used, depending on the size and site of the tumor. The rhinopharyngeal MM reported by Nilles et al was treated with paralateral nasal rhinotomy and showed no signs of relapse at the 6-month follow-up.¹¹ Nofal used the transpalatal approach to remove an MM, but the patient's follow-up was not reported.¹⁴ Tuncel and associates used the midfacial degloving approach with a Le Fort 1 osteotomy followed by radiotherapy, but the patient died 15 months later from a recurrence and lung metastases.¹² In our patient, the tumoral mass was particularly widespread. Therefore, we adopted the infratemporal fossa approach type C as suggested by Fisch²⁰ for tumors in the cranial base and rhinopharynx with involvement of the internal carotid artery. To our knowledge, our patient is the only case of rhinopharyngeal MM treated with this technique. It enabled complete resection of the lesion, and there have been no signs of recurrence during the 5-year follow-up period.

REFERENCES

1. Barnes L, Appel BN, Perez H, El-Attar AM. Myoepitheliomas of the head and neck: case report and review. *J Surg Oncol* 1985;28:21-28
2. Sciubba JJ, Brannon RB. Myoepithelioma of salivary glands: report of 23 cases. *Cancer* 1982;49:562-572
3. Sheldon WH. So-called mixed tumours of the salivary glands. *Arch Pathol* 1943;35:1-20
4. Stromeyer FW, Haggitt RC, Nelson JF, Hardman JM. Myoepithelioma of minor salivary gland origin. *Arch Pathol* 1975;99:242-245
5. 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;109:995-998
6. Chaudhry AP, Satchidanand S, Peer R, Cutler LS. Myoepithelial cell adenoma of the parotid gland: a light and ultrastructural study. *Cancer* 1982;49:288-293
7. Kahn LB, Schoub L. Myoepithelioma of the palate. Histochemical and ultrastructural observations. *Arch Pathol* 1973;95:209-212
8. Seifert G, Sobin LH. The World Health Organization histological classification of salivary gland tumors. A commentary on the second edition. *Cancer* 1992;70:379-385
9. Young J, Barker M, Robertson T, 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;55:230-232
10. Dean A, Sierra R, Alamillos FJ, et al. Malignant myoepithelioma of the salivary glands: clinicopathological and immunohistochemical features. *Br J Oral Maxillofac Surg* 1999;37:64-66
11. Nilles R, Lenartz T, Kaiserling E. Myoepithelial carcinoma of the nasopharynx. Case report and review of the literature. *HNO* 1993;41:396-400
12. Tuncel U, Ergul G, Ozlugedik S, Unal A. Myoepithelial carcinoma in the nasopharynx: an unusual localization. *Yonsei Med J* 2004;45:161-165
13. Nofal F. Spindle cell carcinoma of the nasopharynx. *J Laryngol Otol* 1983;97:1057-1063
14. De Stefani A, Lerda W, Bussi M, Valente G, Cortesina G. Tumori dello spazio parafaringeo. Presentazione di un caso clinico di mioepitelioma a cellule chiare del lobo profondo della ghiandola parotide e revisione della letteratura. *Acta Otorhinolaryngol Ital* 1999;19:276-282
15. 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;7:279-287
16. Torlakovic E, Ames ED, Manivel JC, Stanley MW. Benign and malignant neoplasm of myoepithelial cells: cytologic findings. *Diagn Cytopathol* 1993;9:655-660
17. el-Naggar A, Batsakis JG, Luna MA, Goepfert H, Tortoledo ME. DNA content and proliferative activity of myoepitheliomas. *J Laryngol Otol* 1989;103:1192-1197
18. Ibrahim R, Bird DJ, Sieler MW. Malignant myoepithelioma of the larynx with massive metastatic spread to the liver: an ultrastructural and immunocytochemical study. *Ultrastruct Pathol* 1991;15:69-76
19. Saveria AT, Sloman A, Huvos AG, Klimstra DS. Myoepithelial carcinoma of the salivary glands: a clinicopathologic study of 25 patients. *Am J Surg Pathol* 2000;24:761-774
20. Fisch U. The infratemporal approach for nasopharyngeal tumors. *Laryngoscope* 1983;93:36-44

Commentary

This is a well-written case report of a rare disease in a rare location. The authors ably discuss the clinical and histopathologic presentation that characterizes this rare neoplasm. Because it is un-

likely that a series of such tumors in this location could be developed, a description of this patient's clinical course and the review of the existing related reports represent an especially important addition to our current literature.

Clarence T. Sasaki, M.D.¹