

Recurrent extensive plasmacytoid myoepithelioma of the sinonasal cavity

Amir H. Jafarian, Abbas A. Omid, Nema M. Roshan, Mehdi Montazer, Bahareh Joushan¹

Department of Pathology, Ghaem Hospital, ¹Department of Endodontics, Faculty of Dentistry and Dental Research Center, Mashhad University of Medical Sciences, Mashhad, Iran

Myoepithelioma is an uncommon benign neoplasm that most likely occurs in the parotid gland. Extra-salivary myoepitheliomas are rare, with less than 10 cases reported in the sinonasal cavity. We present a rare case of benign myoepithelioma with recurrent behavior, abundant extensions to adjacent structures, and resistant to treat clinical course, which influenced the patient's quality of life for more than 18 years. Histologic, immunohistochemical, and the potential differential diagnoses are discussed. The patient refused to undertake any more treatments, but after 15 months of follow-up, the lesion did not progress further.

Key words: Iran, myoepithelioma, nasal cavity, neoplasm

INTRODUCTION

The term myoepithelioma was first introduced by Sheldon in 1943.^[1] Myoepitheliomas are generally benign neoplasms composed exclusively of myoepithelial cells. A myoepithelioma classically presents as an asymptomatic mass that slowly enlarges over a long period of time. However, recurrent and malignant ones have also been described.^[2] They are more likely to occur in the parotid gland, but rare cases arising from minor salivary glands are also on record.^[3-5] The authors present an exceptionally rare case of sinonasal myoepithelioma with recurrent and resistant to treat clinical course. To our knowledge and regarding the Medline database, this is the seventh case of its kind until 2012.

CASE REPORT

The patient was a 57-year-old Iranian male living in Dargaz, a small district in north east of Iran. He was admitted to the E.N.T ward, Ghaem Hospital, Mashhad University of Medical Sciences in June 2010. He presented with a 17-year history of recurrent and resistant to treat sinonasal mass. First, at 1993, he accidentally discovered a small painless mass of the

palate, which was excised in an outpatient care facility. After 4 years, the lesion recurred and the patient underwent medial maxillectomy. Thereafter, the patient had five more debulking surgeries.

At the time of admission, the patient was complaining of nasal fullness, hyponasal speech, snoring, oral breathing, and loss of hearing on left side.

On oral examination, the hard palate in the right side had been removed and a nasal mass was obvious through it. Uvula was in the midline. Both nasal cavities were completely filled with the mass. There was no cervical lymphadenopathy. The right tympanic membrane was retracted and the Weber test was shifted to right. All other physical examinations were unremarkable.

Laboratory tests were within normal limits. Family and past medical histories were unremarkable and the patient was non-smoker.

Both computed tomography scan and magnetic resonance imaging demonstrated a huge bilateral lobulated sinonasal mass with extension to cribriform plate and near the dura, medial walls of both orbits, and upper gingival area [Figure 1]. The plain chest x-ray was normal.

The patient underwent a functional endoscopic sinus surgery to debulk the tumor. The removed specimen was stored in 10% buffer formalin and sent to the Pathology Department. The specimen was made by multifragmented irregular-shaped brownish soft tissue with the dimensions of 3.5 × 3 × 2 cm. The whole tissue was processed, cut, and stained with H and E. Microscopically,

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Address for correspondence: Dr. Mehdi Montazer, Department of Pathology, Ghaem Hospital, TaghiAbad Sq., Mashhad, Zip code: 9176699199, Iran. Email: mehdi.montazer@gmail.com

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the specimen showed a hypercellular tumoral tissue composed of plasmacytoid cells with eccentrically located round-ovoid nuclei and acidophilic cytoplasm. The neoplastic cells were arranged in nests, cords, and sheaths intermixed with partially mucoid and hyalinized collagenous stroma. Scattered gland-like structures bordered by neoplastic cells were seen constituting less than 5% of the tissue. No atypia, mitotic activity, necrosis, and vascular-neural invasion were seen. Dispersed foci of spindle cells were also present surrounded by plasmacytoid cells [Figure 2]. A diagnosis of plasmacytoid myoepithelioma was made.

On immunohistochemistry staining, the specimen showed strong and diffuse reactivity for Vimentin, cytokeratin, smooth muscle actin and S-100 confirming the myoepithelial nature of the tumoral cells [Figure 3].

The patient refused to undertake any more treatments. After 15 months of follow-up, the lesion did not progress further.

DISCUSSION

Myoepithelioma is a generally benign neoplasm that constitutes approximately 1–1.5% of all salivary gland neoplasms.^[2] They are more likely to occur in the parotid gland, but rare cases arising from minor salivary glands such as in palate, gingiva, lips, nasopharyngeal space, orbit, and middle ear are also on record.^[3-5] Exceptionally, they can occur in the sinonasal cavity, with only seven reports, including our case, present in the literature.^[6-11]

Histologically, myoepitheliomas are classified as spindle, plasmacytoid, reticular, epithelioid, and clear cell types. The most common is the spindle type, whereas the others are very rare.^[2] All the previously reported sinonasal myoepitheliomas were composed mostly of spindle cells and to a lesser extent of plasmacytoid/epithelioid cells.^[6-11] However, the present case was almost entirely composed of plasmacytoid cells, with dispersed foci of spindle cells.

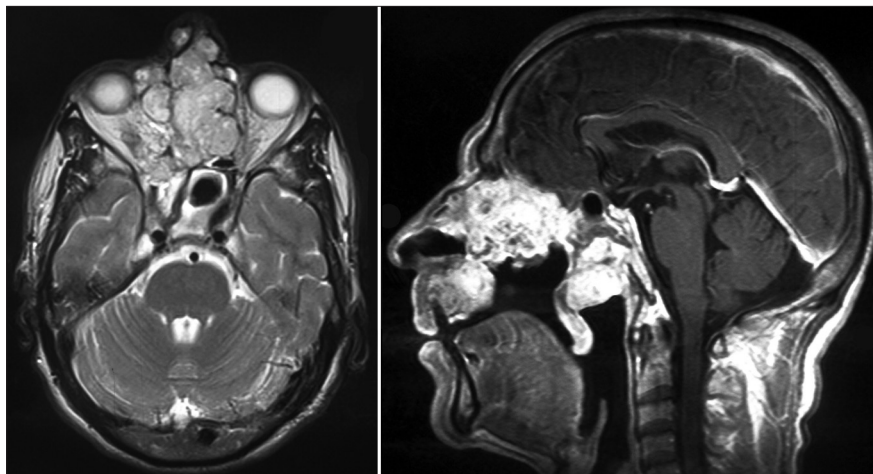


Figure 1: Magnetic resonance imaging. T1-weighted transverse (left) and sagittal (right) planes show a huge bilateral sinonasal mass

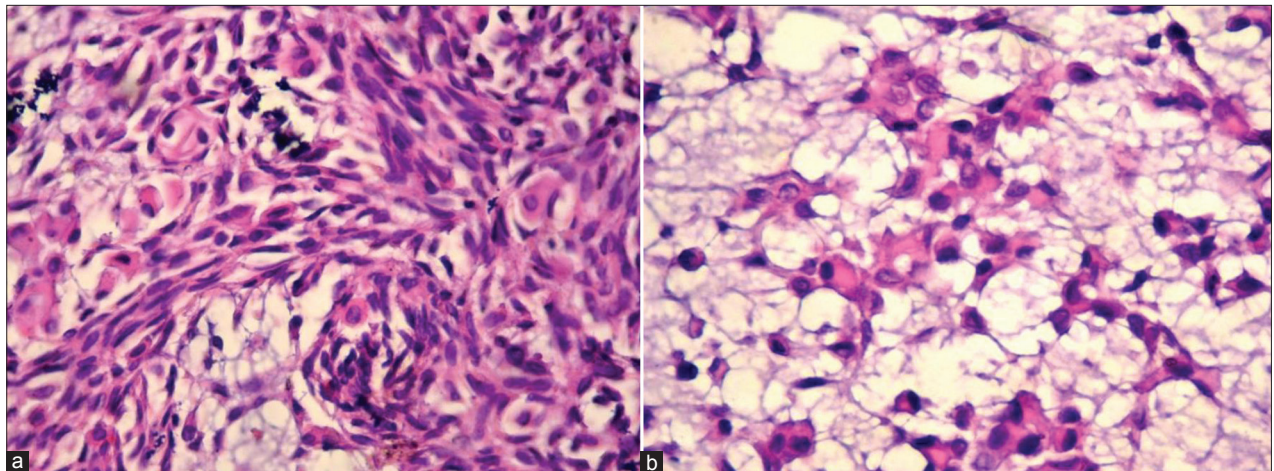


Figure 2: Microscopic view (H and E stain, × 400). (a) A focus of spindle cells. Plasmacytoid cells are also present. (b) Plasmacytoid cells in the background of mucoid material

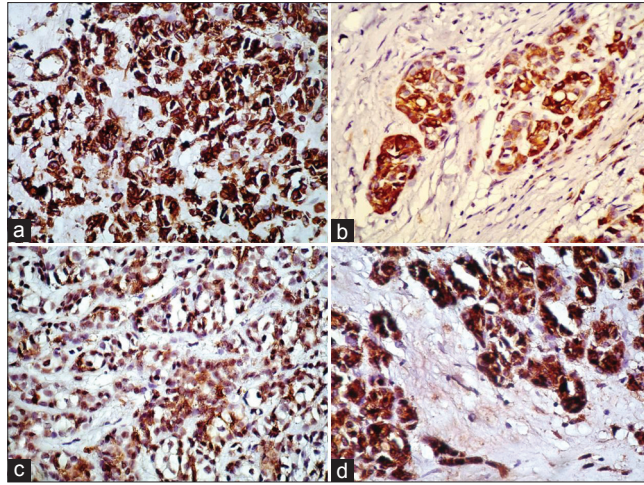


Figure 3: (a) The specimen strongly expresses vimentin, (b) cytokeratin, (c) smooth muscle actin, and S-100 (d) confirming its myoepithelial nature

The most important differential diagnosis of myoepitheliomas includes pleomorphic adenoma. There is a general tendency to put myoepitheliomas at one end of the histologic spectrum that includes pleomorphic adenomas at the other end. This claim is supported by the fact that these two entities are very similar in biologic course, distribution, morphology, and even immunohistochemical aspects. Indeed, the criteria to differentiate these lesions are mainly subjective. World Health Organization proposes that if the neoplasm contains less than 5% of ductal components they must be named myoepithelioma.^[2,5]

Furthermore, several mesenchymal neoplasms such as fibrosarcoma, leiomyoma, schwannoma, and myxoma are in the differential diagnoses of the spindle cell myoepitheliomas, but they all lack the immunoreactivity for cytokeratin.^[6] However, in the presented case, the non-spindle (plasmacytoid) appearance of the neoplastic cells can easily rule out these entities. Moreover, the specimen expressed cytokeratin.

Immunohistochemical examination is a proper technique to facilitate the diagnosis of myoepithelioma. Myoepitheliomas are characteristically immunoreactive for cytokeratins, S-100, Calponin, smooth muscle actin, myosin, vimentin, glial fibrillary acidic protein, and carcinoembryonic antigen.^[6,12] In the presented case, the smooth muscle actin, vimentin, cytokeratin, and S-100 protein were positive.

Conventional myoepitheliomas are not more aggressive than pleomorphic adenomas, but invasive myoepitheliomas that infiltrate the adjacent tissues have been described. In addition, malignant/metastatic ones are also on record. The malignancy is assisted by the infiltrative margins, necrosis, marked pleomorphism, and prominent mitotic activity,^[6,13,14] which were all absent in the presented case. Moreover, high recurrence rates among benign myoepitheliomas have been

described. Recurrence is associated with involved surgical margins at the first excision.^[2] In the present case, the tumor had a benign morphology with a progressive, invasive, and recurrent clinical course, as the tumor has been recurred six times within 17 years. Therefore, the surgical excision must be done with wide healthy margins and an appropriate follow-up is also recommended.

Myoepitheliomas are generally well-circumscribed solid masses that measure less than 3 cm in diameter.^[2] However, in our case, the tumor was a huge bilateral sinonasal mass, which showed prominent extension to cribriform plate, dura, both orbits, and upper gingiva, so that complete surgical removal was practically impossible. To our knowledge, this is the most extensive myoepithelioma of its kind.

Some of the malignant myoepitheliomas have been arisen from pleomorphic adenomas.^[6,14] Regarding our case, the primary lesion had been diagnosed as a myoepithelioma; thus, the further transform of a pleomorphic adenoma seems improbable.

CONCLUSION

We reported the seventh case of myoepithelioma of the sinonasal cavity ever diagnosed regarding the Medline database until 2012. Immunohistochemical investigation may be a useful tool in order to reach to the appropriate diagnosis while a myoepithelioma is in the differential diagnosis of a tumor of the oral/sinonasal cavity. Moreover, complete surgical excision is necessary.

REFERENCES

1. Sheldon WH. So-called mixed tumours of the salivary glands. *Arch Pathol* 1943;35:1-20.
2. Cardesa A, Alos L. Myoepithelioma. In: Barnes L, Eveson JW, Reichart P, Sidransky D, editors. *World Health Organization*

- classification of tumours. Pathology and genetics, head and neck tumours. Lyon: IARC Press; 2005. p. 259-60.
3. Gaio E, Perasole A, Bagatella F. Bilateral myoepithelioma of the nasopharynx: A case report. *Auris Nasus Larynx* 2009;36:496-500.
 4. Herlihy EP, Rubin BP, JianAmadi A. Primary myoepithelioma of the orbit in an infant. *J AAPOS* 2009;13:303-5.
 5. Buchner A, Merrell PW, Carpenter WM. Relative frequency of intra-oral minor salivary gland tumors: A study of 380 cases from northern California and comparison to reports from other parts of the world. *J Oral Pathol Med* 2007;36:207-14.
 6. Sayed SI, Kazi RA, Jagade V, Palav RS, Shinde VV, Pawar PV. A rare myoepithelioma of the sinonasal cavity: Case report. *Cases J* 2008;1:29.
 7. Begin LR, Rochon L, Frenkiel S. Spindle cell myoepithelioma of the nasal cavity. *Am J Surg Pathol* 1991;15:184-90.
 8. Lateef SS, Castillo M, Mukherji SK, Cooper LL. Myoepithelioma of the nasal piriform aperture: CT findings. *AJR Am J Roentgenol* 1999;173:1413-4.
 9. Chang Chao GH, Chung Huang HT, Wul HC. Nasal myoepithelioma: A case report. *Tzu Chi Med J* 1999;11:297-300.
 10. Boscolo-Rizzo P, Da Mosto MC, Marchiori C. Sinonasal spindle-cell myoepithelioma. *Internet J Otorhinolaryngol* 2004;3.
 11. Nakaya K, Oshima T, Watanabe M, Hidaka H, Kikuchi T, Higashi K, *et al.* A case of myoepithelioma of the nasal cavity. *Auris Nasus Larynx* 2010;37:640-3.
 12. Zelaya FC, Rivera DQ, Tapia Vazquez JL, Paez Valencia C, Gaitan Cepeda LA. Plasmacytoid myoepithelioma of the palate. Report of one case and review of the literature. *Med Oral Patol Oral Cir Bucal* 2007;12:e552-5.
 13. Suba Z, Nemeth Z, GyulaiGaal S, Ujpal M, Szende B, Szabo G. Malignant myoepithelioma. Clinicopathological and immunohistochemical characteristics. *Int J Oral Maxillofac Surg* 2003;32:339-41.
 14. Karatzanis AD, Drivas EI, Giannikaki ES, Lachanas VA, Hatzioannou JK, Velegrakis GA. Malignant myoepithelioma arising from recurrent pleomorphic adenoma of the soft palate. *Auris Nasus Larynx* 2005;32:435-7.

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