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Clinical prevalence of myoepithelial carcinoma of the maxillary sinus occurring in north Indian patients

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Abstract
To the best of our knowledge there are very few reports affecting the maxillary sinus in the literature, which makes the maxillary sinus an unusual location and is considered difficult to access for the head and neck surgeon due to the presence of vital structures in the region. As the tumor is so rare, there are no such specific indications and guidelines for its management. Hence the present study was planned to evaluate the myoepithelial carcinoma of the maxillary sinus in North Indian Patients.
The present study was planned in the Department of Dentistry Jay Dental care, Patna, Bihar. Total 10 patients of Histo pathologically confirmed cases of malignancies of nose and Para nasal sinuses are studied with regard to their clinical presentation, radiology, histopathology and treatment protocols. The study was planned from July 2018 to March 2019. All the patients were informed consents. The aim and the objective of the present study were conveyed to them. Approval of the institutional ethical committee was taken prior to conduct of this study.
The data generated from the present study concludes that malignancies of the nose and Para nasal sinuses are an intriguing group of disorders due to the variations in their clinical presentation, histopathological diagnosis, staging and grading. The type of treatment modality also varies depending upon the aforementioned factors. Myoepitheliomas are relatively rare tumors of the salivary glands, with a malignancy potential. As it is a silent neoplasm, they are neither easy to diagnose nor are there any specific protocol/guidelines for its treatment, which makes the prognosis poor.

Keywords: Paranasal sinuses, myoepithelial carcinoma, maxillary sinus, etc

Introduction
The pyramid-shaped maxillary sinus (or antrum of Highmore) is the largest of the paranasal sinuses, and drains into the middle meatus of the nose [1]. It is the largest air sinus in the body. Found in the body of the maxilla, this sinus has three recesses: an alveolar recess pointed inferiorly, bounded by the alveolar process of the maxilla; a zygomatic recess pointed laterally, bounded by the zygomatic bone; and an infraorbital recess pointed superiorly, bounded by the inferior orbital surface of the maxilla. The medial wall is composed primarily of cartilage. The ostia for drainage are located high on the medial wall and open into the semilunar hiatus of the lateral nasal cavity; because of the position of the ostia, gravity cannot drain the maxillary sinus contents when the head is erect (see pathology). The ostium of the maxillary sinus is high up on the medial wall and on average is 2.4 mm in diameter; with a mean volume of about 10 ml [1, 2].
The sinus is lined with mucoperiosteum, with cilia that beat toward the ostia. This membrane is also referred to as the "Schneiderian Membrane", which is histologically a bilaminar membrane with pseudostratified ciliated columnar epithelial cells on the internal (or cavernous) side and periosteum on the osseous side. The size of the sinuses varies in different skulls, and even on the two sides of the same skull [2].
The infraorbital canal usually projects into the cavity as a well-marked ridge extending from the roof to the anterior wall; additional ridges are sometimes seen in the posterior wall of the cavity and are caused by the alveolar canals. The mucous membranes receive their postganglionic parasympathetic nerve innervation for mucous secretion originating from the greater petrosal nerve (a branch of the facial nerve). The superior alveolar (anterior, middle, and posterior) nerves, branches of the maxillary nerve provide sensory innervation.
Myoepithelial carcinoma (malignant myoepithelioma) is a rare salivary gland tumor composed entirely of myoepithelial cells that exhibit a dual epithelial and smooth muscle phenotype. The tumor shows wide morphologic and cytologic diversity in a similar way to its benign counterpart, myoepithelioma, with evidence of malignant change. The malignancy manifests mainly as an infiltrative growth pattern, angiolymphatic and/or perineural invasion, with a propensity for mostly distant metastasis and occasional regional lymph node involvement. Other corroborative evidence of malignancy such as necrosis and increased mitotic activity can also be seen [3].
Originally described by Stromeyer *et al* in 1975 [4], Barnes *et al* renewed interest in this rare entity in 1985 after describing 3 cases of myoepithelial carcinoma (malignant myoepithelioma) in their review of myoepitheliomas of the head and neck [5]. However, it was Dardick *et al*. detailed descriptions of myoepitheliomas that were crucial in furthering the understanding of myoepithelial tumors [6, 7]. Their articles helped to increase the accuracy of describing ensuing reported cases of myoepithelial carcinoma (malignant myoepithelioma). In 1991, myoepithelial carcinoma was added to the second edition of the World Health Organization (WHO) classifications of malignant salivary gland tumors [8].
Many of these tumors arise as a malignant transformation in the setting of a benign pleomorphic adenoma or a benign myoepithelioma [9], and myoepithelial carcinoma (malignant

myoepithelioma) is particularly seen in association with recurrence of these benign tumors [10]. Other such tumors arise de novo [11].

Myoepithelial carcinoma (malignant myoepithelioma) is a rare tumor with a reported incidence of 0.2% of all salivary gland tumors [12]. However, some authors contend that myoepithelial carcinoma (malignant myoepithelioma) may not be as rare as previously suggested [13]. A lack of recognition and/or awareness of its diversity and diagnostic criteria may contribute to the relatively small number of reported cases.

Different series of myoepithelial carcinoma (malignant myoepithelioma) have showed variability in sex distribution with a generally similar age range, although the pediatric age group was not represented in all the relatively large reported series. Myoepithelial tumors are unencapsulated soft to firm masses with infiltrative margins, although some may appear well delineated grossly. The size ranges from 2-20 cm [14]. A multinodular appearance can be appreciated on gross examination in many of these tumors. The cut surface may show focal hemorrhage, cystic degeneration, and necrosis. Myoepithelial carcinomas (malignant myoepitheliomas) are unencapsulated and usually multinodular in appearance, with infiltrative borders [15]. The nodules vary in size with intervening thin fibrous septae, and they can exhibit central nodular necrosis. Other less-common growth patterns include diffuse solid, trabecular, and reticular infiltrating patterns [16].

Like all tumors with myoepithelial component, myoepithelial carcinomas (malignant myoepitheliomas) exhibit a wide morphologic and cytologic diversity that is based on 4 major types of cells similar to those seen in benign myoepithelioma: spindle, plasmacytoid (hyaline), epithelioid, and clear cell types. Many myoepithelial carcinomas (malignant myoepitheliomas) exhibit more than one cell type, but even in these, one cell type usually predominates. Other cell morphologies such as stellatelike or vacuolated cells have also been described [16], as well as some tumors that exhibit an oncocytic morphology. In these malignant neoplasms, however, the cell type does not appear to significantly influence a patient's survival [17].

The epithelioid cells are large polygonal cells with central ovoid or round nuclei and eosinophilic or amphophilic, sometimes focally clear cytoplasm (see the following image); these can form pseudoacini or pseudoglandular structures (not true glands) [18]. Myoepithelial carcinoma (malignant myoepithelioma) exhibits a propensity for distant metastasis more than for regional lymph node metastasis, with an ability by most tumors for extensive local growth, infiltration, and destruction. Distant metastasis has been seen to the lungs (most commonly), bone, liver, peritoneum, pleura, kidneys, brain, and skin. A study that looked to provide a better characterization of myoepithelial carcinoma and its prognostic factors reported that myoepithelial carcinoma is an aggressive tumor that is associated with a high rate of distant metastasis and compared with de novo myoepithelial carcinoma, carcinoma ex-pleomorphic adenoma correlates with worse clinical outcome [19].

It can be gleaned from the studies above that to the best evidence provided, and with around 50% to over 65% survival from cases that were followed up in these reported series, myoepithelial carcinoma (malignant myoepithelioma) is probably best considered a tumor with high-grade potential and unpredictable biologic behaviour. Careful patient follow-up and staging is therefore essential for better characterization and understanding of this tumor's behaviour in the future.

To the best of our knowledge there are very few reports affecting the maxillary sinus in the literature, which makes the maxillary sinus an unusual location and is considered difficult to access for the head and neck surgeon due to the presence of vital structures in the region. As the tumor is so rare, there are no such specific indications and guidelines for its management. Hence the present study was planned to evaluate the myoepithelial carcinoma of the maxillary sinus in North Indian Patients.

Methodology

The present study was planned in the Department of Dentistry Jay Dental Care, Patna, Bihar. Total 10 patients of Histopatho logically confirmed cases of malignancies of nose and Para nasal sinuses are studied with regard to their clinical presentation, radiology, histopathology and treatment protocols. The study was planned from July 2018 to March 2019. All the patients were informed consents. The aim and the objective of the present study were conveyed to them. Approval of the institutional ethical committee was taken prior to conduct of this study.

Following was the inclusion and exclusion criteria for the present study.

Inclusion Criteria: 1. All Histo pathologically confirmed cases of malignant neoplasms of nose and Para nasal sinuses irrespective of sex were considered for study.

2. All patients who were immunologically competent.

Exclusion Criteria: 1. Paediatric malignancies are excluded.

2. All immune compromised patients are excluded from the study.

Diagnostic nasal endoscopy (DNE) revealed a pink fleshy mass seen in the left nasal cavity arising from the left middle meatus. Examination of the oral cavity revealed a smooth bulge measuring 2× 2 centimetres seen on the soft palate on the left side, which was firm in consistency, with no tenderness on palpation. CT scan showed a heterogeneous soft tissue density in the left maxillary sinus and the nasal cavity with erosion of the floor of the orbit and inferior wall of the maxilla and extending into the left infra temporal & Pterygo-palatine fossa. Biopsy was taken with the help of an endoscope from the mass in the left nasal cavity. Histopathology revealed low grade Adenoid cystic carcinoma of the left maxillary sinus.

Results & Discussion

Myoepithelial carcinoma, known as malignant myoepithelioma, is the malignant counterpart of myoepithelioma. Malignant myopithelioma has been added to the second edition of the World Health Organization's histological classification of salivary gland tumor [20]. Manuel *et al*. had previously proposed a classification system comprising de novo invasive or non-invasive pleomorphic carcinoma, invasive or noninvasive carcinoma with no pleomorphic adenoma, and true malignant mixed tumor (carcinosarcoma), which may arise alone or from a preexisting pleomorphic adenoma [21]. Myoepithelial carcinomas of the salivary gland should be classified as high-grade malignancies. Overall, the prognosis of

myoepithelial carcinoma is poor [22].

Epithelial Myoepithelial Carcinoma (EMC) is a low-grade malignancy, only rarely have high-grade or dedifferentiated EMC cases been reported. It has been observed that some morphologically low-grade myoepithelial carcinomas behave aggressively [23].

It is rare tumor and a diagnostic dilemma. It should be considered in cases showing dual tumor cell population with clear cell change in histopathology. Common differential diagnosis includes metastatic renal cell carcinoma, acinic cell carcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma, oncocytoma, and sebaceous carcinoma [24]. Renal cell carcinoma can be distinguished by presence of multiple vessels and CD10 positivity. Acinic cell carcinoma can be excluded by immunohistochemistry since it is negative for all markers. Mucoepidermoid carcinoma shows distinct mucoid areas. EMC must be distinguished from adenoid cystic carcinoma, and these two forms can be described together as the so-called "hybrid" carcinoma, when coexistent [25]. Sebaceous carcinoma is positive for EMA and cam5.2.Oncocytoma can be distinguished by the presence of abundant eosinophilic granular cytoplasm.

Table 1: Age of Cases and No. of Cases

Age in years	No. of cases
0-10	0
11-20	1
21-30	0
31-40	3
41-50	2
51-60	3
61-70	1
Total	10

Table 2: Frequency of presenting symptoms of sinonasal epithelial-myoepithelial carcinoma

Symptoms	No. of cases
Epistaxis	6
Nasal obstruction	7
Facial Swelling	4
Headache	1
Epiphora	0
Hyposmia	0
Vision change	0

Table 3: Site of tumor origin

Symptoms	No. of cases
Inferior turbinate	2
Maxillary Sinus	3
Nasal septum	1
Anterior ethmoid	1
Nasal cavity unspecified	2
Unknown	1
Total	10

The differential diagnoses of myoepithelial carcinoma encompasses a wide variety of tumors including myoepithelioma, epithelial myoepithelial carcinoma, clear cell adenocarcinoma, synovial sarcoma, malignant peripheral nerve sheath tumor, leiomyoma, plasmacytoma, malignant melanoma and many more owing to its morphological heterogeneity [26]. Myoepithelial carcinomas may demonstrate intacytoplasmic glycogen on periodic acid-Schiff (PAS) staining which is sensitive to diastase, particularly in the clear cell subtype [27]. Immunohistochemically, myoepithelial carcinomas are positive for cytokeratin including Cam 5.2, AE1:AE3 and 34β12, vimentin and S-100. Smooth muscle markers including calponin, smooth muscle actin (SMA), muscle-specific actin (MSA), smooth muscle myosin (SMM), caldesmon, glial fibrillary acidic protein (GFAP), CK14, CD10, p63 and Bcl2 may be expressed in a proportion of cases and calponin appears to be most sensitive [28, 29]. Diagnosis requires coexpression of cytokeratin and one of the myoepithelial markers [30]. Carcinoembryonic antigen (CEA; marker for luminal differentiation), epithelial membrane anitgen (EMA), CK7, desmin and CD57 are usually negative [5, 9, 10, 11]. P53 overexpression may indicate a worse outcome [10]. Myoepithelial carcinoma is composed of modified myoepithelial cells which may not show complete immunohistochemical expression profile of non-neoplastic myoepithelial cells. In our case, the tumor cells showed immunohistochemical expression of S100, pancytokeratin and focal SMA.

According to some authors, myoepithelial carcinomas arising de novo may be more aggressive tumors as compared to those arising in recurrent pleomorphic adenomas [31]. Myoepithelial carcinoma arising in pleomophic adenoma may be an under reported category, probably due to difficulty in diagnosing the myoepithelial component especially in the absence of marked cytological atypia, mitoses or necrosis; and the inconspicuous nature of the coexisting pleomorphic adenoma [32]. Evidence of a preexisting pleomorphic adenoma at the same site may be crucial in reaching accurate diagnosis, which is essential for optimal management of the patient considering the risk of recurrence and metastases.

Conclusion

The data generated from the present study concludes that malignancies of the nose and Para nasal sinuses are an intriguing group of disorders due to the variations in their clinical presentation, histopathological diagnosis, staging and grading. The type of treatment modality also varies depending upon the aforementioned factors. Myoepitheliomas are relatively rare tumors of the salivary glands, with a malignancy potential. As it is a silent neoplasm, they are neither easy to diagnose nor are there any specific protocol/guidelines for its treatment, which makes the prognosis poor.

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