Nasopharyngeal Mucoepidermoid Carcinoma - A Common Entity at an **Uncommon Location**

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ABSTRACT

Mucoepidermoid carcinomas mostly occur in the major salivary glands, the minor salivary glands of oral cavity and in the lacrimal glands. These tumours rarely occur in the sino-nasal tract. When they occur in the sino-nasal tract, the most frequent site is the maxillary antrum, followed by the nasal cavity, the nasopharynx and the ethmoidal sinuses. As per review of literature, nasopharyngeal mucoepidermoid carcinomas account for 0.6% of salivary gland tumours and 4.8% of mucoepidermoid carcinomas. Extensive literature search revealed 21 cases of nasopharyngeal mucoepidermoid carcinomas reported till date. These cases showed an age incidence ranging from 20 to 60 years with a female preponderance.

In contrast to nasopharyngeal carcinomas, these tumours show low positivity rates for Ebstein-Barr virus serological test. Histochemical positivity for mucin may be demonstrated in the glandular and mucinous components of these tumours. High grade mucoepidermoid carcinoma of nasopharynx is treated with surgical excision combined with radiotherapy and is associated with poor survival. Therefore, early diagnosis and prompt treatment are of utmost importance.

This case report highlights the rare occurrence of a high grade nasopharyngeal muco-epidermoid carcinoma in a 70-year-old male and is presented for its unusual occurrence in the nasopharynx which is the most infrequent location for this lesion.

Keywords: Mucoepidermoid Carcinoma, Nasopharynx, Ebstein-barr virus

CASE SUMMARY

A 70-year-old male presented with a swelling in the left lateral side of the neck of 1 month's duration. The swelling was progressive in nature and not associated with pain or fever initially, but subsequently the patient developed pain and difficulty in swallowing. The patient was a chronic smoker since 20 years who smoked 5 to 6 cigarettes per day.

Local Examination

Inspection: A solitary left cervical swelling at level II measuring about 6x4 cm with smooth surface was seen.

Palpation: Size-6x4cms, Borders-well made out, Consistencyhard, Mobile.

Fine needle aspiration cytology of the left cervical lymph node showed metastatic carcinomatous deposits.

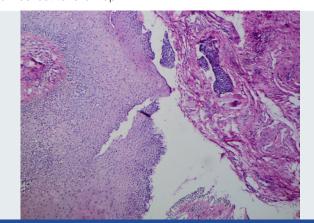
Indirect laryngoscopy revealed an ulcero-proliferative growth in the nasopharynx. The extent of the growth could not be made out.

A clinical diagnosis of nasopharyngeal malignancy was offered and the punch biopsy from the ulcero-proliferative growth in the nasopharynx was submitted for histopathological examination.

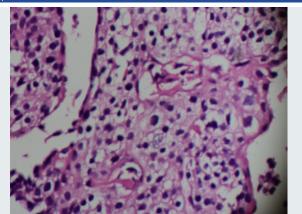
Microscopic examination revealed an invasive subepithelial malignant tumour composed of lobules of tumour tissue with an admixture of squamoid, intermediate and clear cells [Table/Fig-1]. The squamoid component showed marked pleomorphism and a significant number of atypical mitoses. The overlying hyperplastic squamous epithelium showed focal areas of moderate dysplasia [Table/Fig-2].

Special staining with Per-iodic acid Schiff demonstrated focal positivity within the clear cells [Table/Fig-3]. With these findings, a histopathological diagnosis of high grade mucoepidermoid carcinoma of the nasopharynx was arrived at.

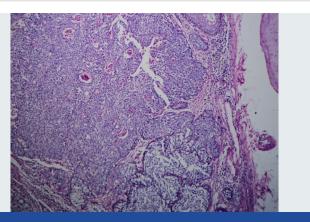
By correlating the fine needle aspiration cytology findings in the left cervical lymph node and the histopathological findings in the nasopharyngeal growth, a final diagnosis of nasopharyngeal mucoepidermoid carcinoma with metastatic deposits in the left cervical lymph node was signed out. Following this report, the patient was immediately referred to a higher centre for further management and was lost to follow up.



[Table/Fig-1]: 10x, H & E - shows stratified squamous epithelium, and



[Table/Fig-2]: 40x, H & E - shows tumour islands composed of squamoid, intermediate and clear tumour cells



[Table/Fig-3]: 40x, PAS - shows focal positivity within clear cells

DISCUSSION

Malignant tumours of the nasal cavity are relatively rare [1]. Only 21 cases are reported till date [2]. The differential diagnosis of malignancies in this area most commonly include Squamous cell carcinoma, Adenocarcinoma, Undifferentiated carcinoma, Melanoma, Lymphoma and very rarely, a Mucoepidermoid carcinoma. Squamous cell carcinomas have been observed to be the most predominant cancers of nasal cavity and account for about 1.7% of nasopharyngeal cancers [3].

Most of the sinonasal tract adenomatous tumours including mucoepidermoid carcinomas are known to originate from the surface mucosa [4].

Nasopharyngeal mucoepidermoid carcinomas and Squamous cell carcinomas may be preceded by mucosal dysplasia or carcinoma in situ. Minor trauma and chronic irritation have been implicated in the etiology of sinonasal tract cancers, besides the sporadic causes like Vitamin A deficiency, radiation exposure and occupational exposure to wool dust, industrial toxins, nickel, chrome, leather, textiles and clothing. Our patient had no history suggestive of any of these factors, but he was a chronic smoker. However, there was no evidence in literature that cigarette smoking is a causative factor.

The presenting signs and symptoms in a patient with nasopharyngeal mucoepidermoid carcinoma usually include pain, nasal obstruction, epistaxis, diplopia and a subcutaneous mass if there was bony erosion. In contrast, our patient presented with dysphagia which is an unusual presenting feature.

The tumour is graded based on morphologic features such as

- 1. Proportion of cystic and solid elements
- 2. Neural invasion
- Necrosis

- 4. Mitotic rate
- 5. Anaplasia [5].

A point value is assigned to each of these parameters and the total score heips to determine the tumour grade as low, intermediate or high. Based on these parameters and scoring, the tumour grade in the present case was assigned as high. An overall ten year cure rate for mucoepidermoid carcinoma in the head and neck as 90% for low grade and 42% for high grade tumours have been reported by Spiro et al. Bone invasion, age beyond 60 years, pain, positive cervical nodes and facial nerve palsy have been implicated as other poor predictive factors besides high grade [6].

The treatment should be based on tumour grade and extent of invasion. Low grade mucoepidermoid carcinomas respond well to surgical resection alone. Total resection and post-operative radiation are recommended for intermediate and high grade tumours. Our patient was referred to a higher center for management and hence lost to follow up.

Since nasal cavity is an unusual location for mucoepidermoid carcinomas, one may overlook its possibility in this location. Besides this, the affected patient may present with signs and symptoms typical of chronic sinusitis such as nasal obstruction and epistaxis which may mislead the clinician. Although diagnosis of a low or intermediate grade carcinoma poses no diagnostic difficulties to the pathologist, special mucin stains may have to be resorted to differentiate a high grade mucoepidermoid carcinoma from squamous cell carcinoma.

CONCLUSION

Since high grade nasopharyngeal carcinomas are associated with poor survival even with aggressive measures, a timely and prompt diagnosis, accurate grading and early treatment are absolutely necessary.

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