

Myoepithelial Carcinoma Arising In Pleomorphic Adenoma of Palatal Minor Salivary Gland: Report of a Rare Case

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Abstract

Salivary gland neoplasms composed exclusively or predominantly of myoepithelial cells are relatively uncommon and constitute 1 % of all salivary gland tumours. Most of them behave in a benign fashion and are designated as myoepithelioma. The malignant counterpart is known as myoepithelial carcinoma or malignant myoepithelioma, which is quite rare. This report presents about a rare case of a 45 year old female who had a painless swelling in the left posterior palatal region since childhood with a sudden increase in size since 2 months. Pre-operative biopsy failed to reveal any malignant changes. The mass was surgically excised and thorough evaluation revealed a myoepithelial carcinoma arising in pleomorphic adenoma of palatal minor salivary gland.

Key Words: Carcinoma ex pleomorphic adenoma, Myoepithelial carcinoma, Salivary gland tumors.

Introduction

Pleomorphic adenoma (PA) is the most common benign tumour arising from the salivary glands accounting for approximately 52.7% of all salivary gland neoplasms [1]. The most common intraoral site of its occurrence is palate [2,3]. Approximately 25% of pleomorphic adenomas undergo malignant transformation. Different patterns of malignant changes can occur of which carcinoma ex pleomorphic adenoma (CXPA) is the most common form. The two other forms are true malignant mixed tumour (carcinosarcoma) and metastasizing pleomorphic adenoma. CXPA is defined as a carcinoma that arises in the epithelial and/or myoepithelial component of a pleomorphic adenoma. In most of the instances (75%), the luminal epithelial cells undergo malignant change. In 19% of the cases, there is a dual epithelial-myoepithelial differentiated carcinoma. Pure myoepithelial change is rare and seen only in 6 % of the cases [4]. This report presents a rare case of myoepithelial carcinoma arising in pleomorphic adenoma of palatal minor salivary gland.

Case Report

A 45 year old otherwise healthy female reported to Department of Oral Medicine and Radiology with a complaint of painless swelling in the left posterior palatal region since childhood. There was a sudden increase in the size of swelling in past 2 months. Patient had altered sensation in the region of swelling, but no history of nasal blockage or bleeding. Medical history was non-contributory. General physical examination and extraoral examination revealed no abnormality. Intraoral examination revealed, a single lobulated swelling measuring 4x5 cm on the left side of palate, anteroposteriorly extending from region of upper left second premolar to third molar, distally crossing junction of hard and soft palate. Mediolaterally, extending from midline of palate to gingival margin. Mucosa overlying swelling was tense and erythematous but intact (Figure 1). On palpation, the swelling was non-tender, firm in consistency, slightly compressible but not reducible, fluctuant or pulsatile. The adjacent teeth (maxillary left molars) were vital and demonstrated grade II mobility. Presence of long standing swelling and the location were in favour of a benign



Figure 1. Intraoral photograph showing a single lobulated swelling on the left side of palate.

salivary gland neoplasm such as pleomorphic adenoma. But the history of sudden increase in the size of swelling aroused the suspicion of a malignant transformation within the benign lesion. Hence, our differential diagnosis included carcinoma ex pleomorphic adenoma, adenoid cystic carcinoma, mucoepidermoid carcinoma and adenocarcinoma. Panoramic radiograph showed break in continuity of left maxillary sinus floor & absence of radiopaque shadow of hard palate on left side (Figure 2). In CT scan, a heterogenous hypodense area was seen in left maxillary sinus region destroying hard palate, alveolar process of maxilla, inferior turbinate and medial wall of left maxillary sinus (Figure 3). Posterolaterally, the mass was infiltrating into lateral pterygoid muscle. Despite the clinical suspicion of a malignant process, pre-operative biopsy failed to reveal any malignant changes and lesion showed only mild cytological atypia. Considering aggressive nature of the growth left partial maxillectomy with extraction of all the maxillary molars on left side was done. A thorough evaluation of the resected tumour sections showed central areas of myxohyaline and myxochondroid matrix with peripheral solid nests of epithelial and myoepithelial cells, suggesting a pleomorphic adenoma. But focal areas of necrosis, increased mitotic activity, clear cells with anisonucleosis and hyperchromasia, coupled with infiltration of bone suggested

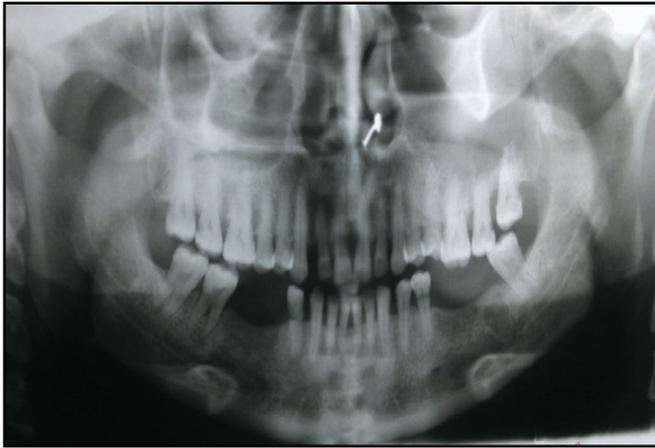


Figure 2. Panoramic radiograph showing break in continuity of left maxillary sinus floor & Absence of radiopaque shadow of hard palate on left side.

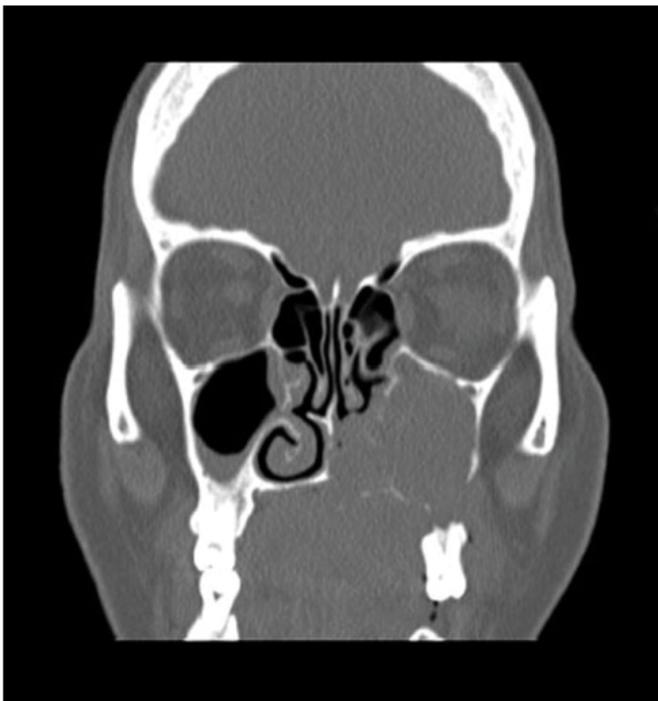


Figure 3 (A, B). CT Scan showing a heterogeneous hypodense area in left maxillary sinus region.

myoepithelial carcinoma arising in a pleomorphic adenoma (Figure 4 A,4B,4C). Immunohistochemical staining showed Ki 67 labelling index of 21 % which was more in favour of an aggressive entity (Figure 4D). Hence, a final diagnosis of myoepithelial carcinoma arising in pleomorphic adenoma was given. After surgical excision, obturator was fabricated to cover the palatal defect and post-operative adjuvant radiation therapy was given for a period of 6 weeks. Patient was on regular follow up for a period of 2 years with no recurrence.

Discussion

Carcinoma ex-pleomorphic adenoma (CXPA) is a rare, aggressive and poorly understood malignancy that usually develops in a primary or recurrent pleomorphic adenoma [5]. The pathogenesis has not been well understood. CXPA usually occurs in the 6th–8th decades of life [6]. The clinical presentation of CXPA may be similar to that of PAs and the most common indication of malignant transformation is a sudden increase in the size.

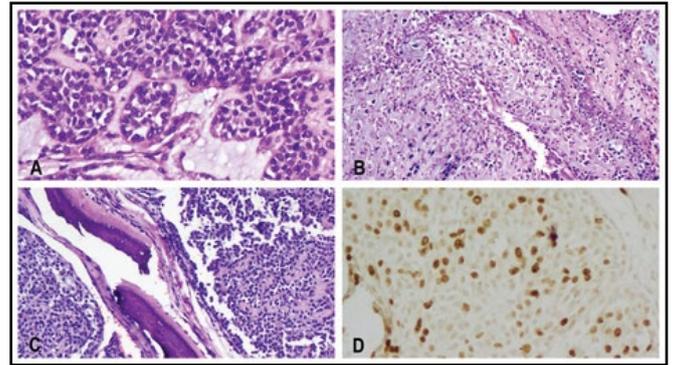


Figure 4 (A) Central areas of myxohyaline and myxochondroid matrix with peripheral solid nests of epithelial & myoepithelial cells, suggesting a pleomorphic adenoma (H & E X 200) (B) Necrotic Areas In Chondroid Foci In Tumour (H & E X 40) (C) Tumour Infiltration of Bone (H & E X 100) (D) Ki 67 Staining which was in favour of an aggressive entity (20X).

Salivary gland neoplasms composed exclusively or predominantly of myoepithelial cells are relatively uncommon and constitute 1 % of all salivary gland tumours. Myoepithelial salivary gland tumor was first identified as a distinct neoplastic entity, in 1943 by Sheldon [7]. The relatively recently recognized and rarely encountered malignant myoepithelial tumor (i.e., myoepithelial carcinoma) was first described by Stromeyer et al. [8] in 1975 and is one of the several new entities included in the updated classification of salivary gland neoplasms by the World Health Organisation in 1991 [9]. Myoepithelial carcinoma may occasionally develop in a pre-existing pleomorphic adenoma, but most often arises *de novo* [10]. Saveria et al. [11] reviewed 25 cases of myoepithelial carcinoma out of which in one case primary tumor site was palate. Some of these carcinomas originated *de novo*, whereas others were classified myoepithelial carcinoma ex pleomorphic adenoma or as arising in benign myoepithelioma. Demasi et al. [12] had done a study in which out of 16 cases of CXPA, 75 % were only epithelial (luminal) malignancy. The study suggested that CXPA with only epithelial differentiation were limited to the major salivary glands. CXPA with myoepithelial differentiation can be further subdivided into those that exhibit both epithelial and myoepithelial malignancy and those with exclusive myoepithelial malignancy. Those with exclusive myoepithelial malignancy are quite rare.

In many cases preoperative biopsy may not be beneficial in revealing the underlying nature of the pathology. Similar problem was encountered while diagnosing our case as the incisional biopsy from tumor site showed only mild cytological atypia. Said et al. [13] had discussed the difficulties that may be encountered in attempting to diagnose these rare myoepithelial lesions by means of small biopsy specimens. Four cell types are identified in myoepithelial salivary gland neoplasms: plasmacytoid, spindle, epithelioid and clear cell types. Of these, the clear cell type is the least frequently encountered [14,15]. In our case, clear cells with anisonucleosis and hyperchromasia were noticed which were in favour of a clear cell variant. Various studies have been done to check the role of Ki-67 in the diagnosis of salivary gland tumours [16,17]. Nagao et al. [18] conducted a study in which the Ki-67 labelling index in myoepithelial carcinoma and benign myoepithelioma were compared and a Ki-67

labelling index of greater than 10% was considered diagnostic of myoepithelial carcinoma. Even in our case the labelling index was quite high and was more in favour of an aggressive neoplasm.

Currently, surgery and post-operative adjuvant radiation therapy are accepted to improve local tumour control and increased survival. Post-operative adjuvant radiation could effectively eradicate residual deposits of microscopic disease [19]. They have high recurrence and metastatic rates, which vary from 25% to 75%. Metastatic lesions most frequently occur in regional lymph nodes, and some of them are seen in lung and bone. Recurrence and regional and distant metastases are predictive of extremely poor prognosis. In one series, disease-specific survival was 45% at 3 years and 37%

at 5 years [20]. However, median survival was 27% at 1 year after detection of any type of progression and 5% at 3 years after detection of distant metastasis.

Conclusion

Clinical presentation of CXPA may be similar to that of pleomorphic adenoma and the most common indication of malignant transformation is a sudden increase in the size. Preoperative biopsy may not be beneficial in revealing the underlying nature of the pathology. Currently, surgery and post-operative adjuvant radiation therapy remains as the modality of choice for most cases. Regular follow-up of the patient with repeated scans and radiographs are advised to prevent recurrence or metastasis.

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