

# Carcinoma ex Pleomorphic Adenoma of Right Hard Palate — A Case Report

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## Abstract

Carcinoma ex pleomorphic adenoma (CXPA) is a rare malignancy of salivary glands. This type of tumor usually arises in major salivary glands, most commonly in parotid gland, which is followed by submandibular gland. CXPA arising in minor salivary glands is extremely rare. CXPA is very difficult to identify before surgical excision for pathologic examination. It is because the clinical features of many cases are similar to those of pleomorphic adenomas (PA). Pathological examination is still the gold standard for diagnosis that—based on the identification of both components of pleomorphic adenoma and carcinoma in the tumor entities. Surgical ablation combined radiation therapy are still the major treatment modalities of CXPA.

Here we presented a case of 45 y/o male who had a painless mass with central ulcer on right palatal mucosa noted for several months. The tumor was excised with 5 mm safe margin by transoral approach. There was no recurrence during the one year follow-up period.

**Key words:** Carcinoma ex pleomorphic adenoma, Minor salivary gland.

## Introduction

Carcinoma ex pleomorphic adenoma (CXPA) is a rare malignancy of head and neck region, in particular, for those in the minor salivary glands. It accounts only 3.6% of all salivary gland tumors and 11.6% of all malignant neoplasms of salivary gland.<sup>1</sup> This type of tumor usually arises in major salivary glands, most commonly in

parotid gland, which is followed by submandibular gland. CXPA arising in minor salivary glands is even rare. It is difficult to distinguish them from pleomorphic adenoma. No matter what clinical features presented clinically, incisional biopsy with thorough image study is indicated for presurgical evaluation. Here we presented a case of carcinoma ex pleomorphic adenoma occurred at right palatal mucosa, and some literatures were

reviewed.

## Case Report

A 45 y/o male patient had an ulcerative mass at right palatal mucosa noted for a month. He had no trauma history of this area. He has been a hepatitis B carrier. Other than that no other specific findings nor family history was mentioned.

Since April 2014 a painless mass was found at his right palatal mucosa. He did not seek for further treatment until surface ulcer developed in May 2014. Oral examination showed: (1) A dome-shaped mass with central ulcer at right posterior palatal mucosa, with 2.0 x 1.8 cm in size and rubbery texture (Fig. 1) (2) Another small bony hard swelling was noted just left to the ulcerative lesion with size of 1 x 0.5 cm.

Under the suspicion of malignancy, biopsy was done and the pathologic report showed pleomorphic adenoma. Magnetic resonance imaging revealed a mass lesion about 2.3 cm was noted of right hard palate (Fig. 2) with intact palatal bone. Several small bilateral cervical lymph nodes was found at level II. Bone scan was done and no abnormal bony uptake suggested no local invasion or distant metastasis to bone for this case (Fig. 3). All the lab examination was within normal range. The tentative diagnosis was pleomorphic adenoma of right hard palate and torus palatines of central hard palate.

We then resected the tumor and the torus firm mass trans-orally with 0.5 cm safe margin under general anesthesia (Fig. 4). Smooth and intact bone surface with mild depression noted of the remained palatal bone. We smoothed the bone surface of the surgical area with bur trimming. The surgical defect was covered by

surgical stent to compress Alloderm stably over raw bone surface.

The pathological examination showed that the tumor majorly composed of ductal and myoepithelial cells with myxoid and hyalinized stroma. Focal malignant transformation with infiltrating growth pattern are noted with the tumor mass (Fig. 5). The tumor margins showed free of tumor cells except basal margin which contact the palatal bone. The special immunohistochemical (IHC) staining, the tumor cells were positive for CK7, p63, calponin, E-cadherin and S-100. The tumor was originated from myoepithelial cells. The final pathological diagnosis was Carcinoma ex pleomorphic adenoma, low grade, pT1NxM0 stage I.

Due to positive of tumor cells of basal margin, further operation or radiotherapy was indicated. The case was discussed by our institute's multidisciplinary head and neck tumor board, and the consensus favored post-operative radiotherapy. However the patient didn't accept this treatment plan. In order to further evaluate the condition of regional or distant metastasis, positron emission tomography/computed tomography (PET/CT) scan was arranged at one month after the operation and the image revealed no evidence of distant metastasis. Further MR image was arranged at two months after operation revealed no residual tumor. The patient kept his routine OPD follow up. After one year regular follow-up after initial treatment. No local recurrence was noted (Fig. 6).

## Discussion

Carcinoma ex pleomorphic adenoma (CXPA) is defined as a carcinoma derived from pre-existed pleomorphic adenoma. It accounts for

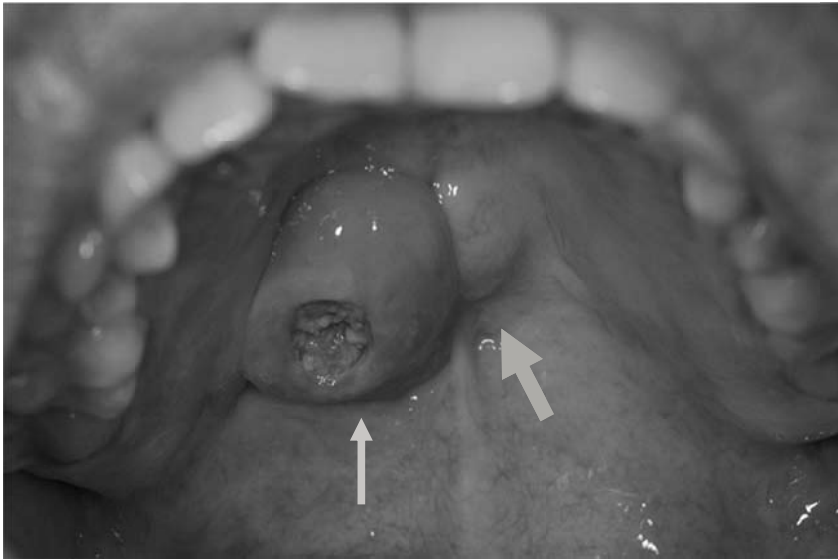


Fig. 1. Swelling mass with central ulceration of right palatal mucosa was noted (red arrow) with another bony hard swelling with intact and smooth mucosa surface at midline of palate (blue arrow).

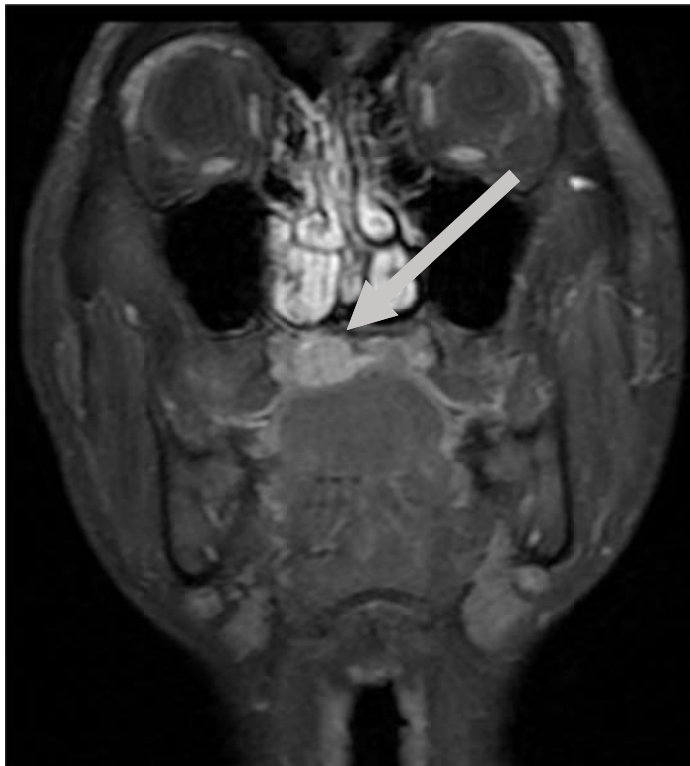


Fig. 2. A tumor mass with enhancing density about 2 x 2 cm noted at right hard palate on the coronal section of MR image.

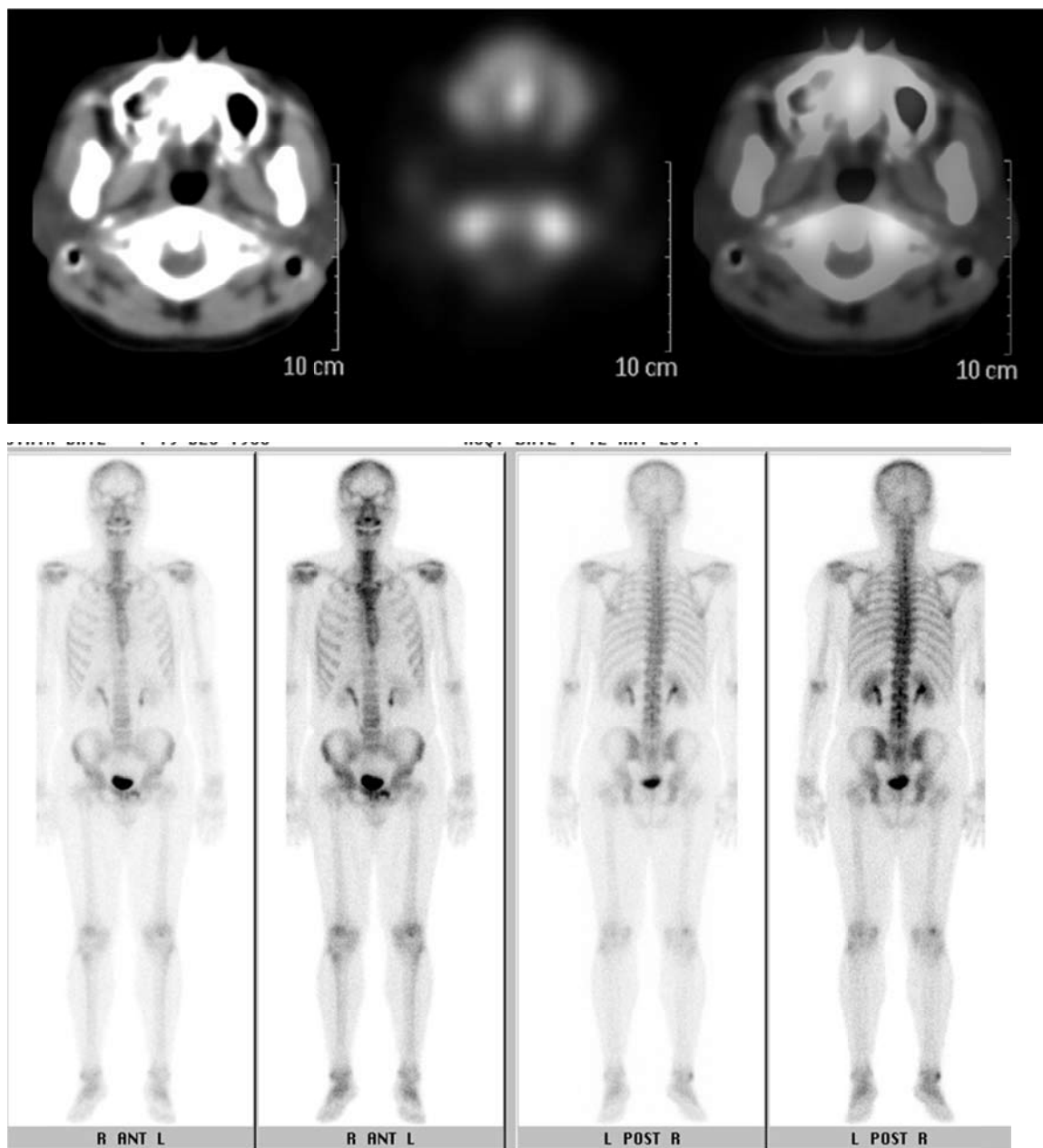


Fig. 3. No abnormal bony uptake that suggests local invasion of palate on the whole body bone scan.

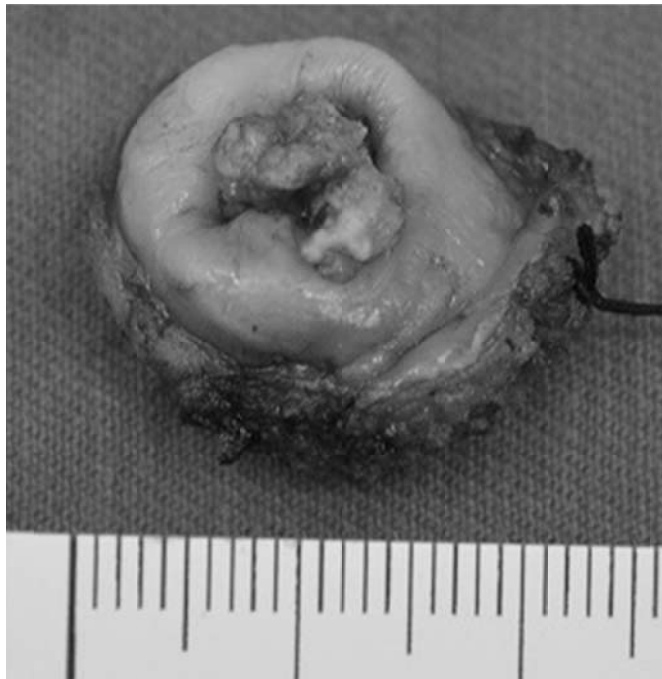


Fig. 4. The specimen of the tumor was excised totally by operation with 1.7 cm in diameter and rubbery texture. Some sandy like material was pulled out from the ulcerative part of the tumor.

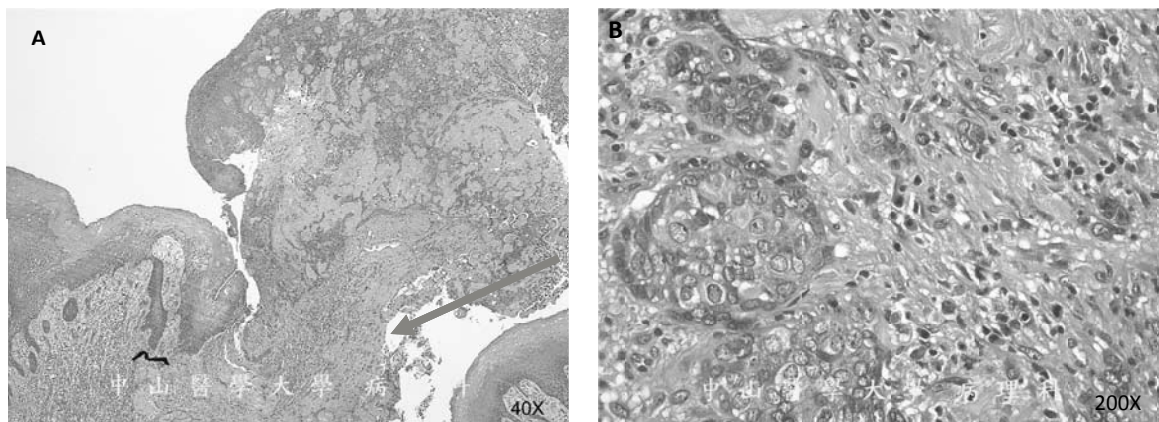


Fig. 5. (A) Hematoxylin and eosin-stained section of surgical specimen showing tumor cells spread out from the normal mucosa. (B) Ductal and myoepithelial cells with myxoid and hyalinized stroma. Focal malignant transformation and infiltrate growth pattern also noted.



Fig. 6. Intraoral picture of post-operative follow-up for more than a year, the mucosa of right hard and soft palate was intact without recurrence.

approximately 3.6% of all salivary gland neoplasms and 11.6% of all malignant salivary gland neoplasms.<sup>1</sup> Majority of CXPA arises in major salivary glands, most in parotid gland followed by submandibular gland. Those in minor salivary glands are extremely rare. From 2000 till now, only 83 cases has been reported. 63 of them occurred at palate (Table. 1). The onset of CXPA is usually at the age of sixth to eighth. Clinically usually presented as a painless mass that is very similar to pleomorphic adenomas (PA). The risk for malignant change of this tumor increases with the existing duration of a mixed tumor. Patients with this disease often aware of the existence of benign tumors for many years and experienced recent rapid growth of their tumors that are associated with symptom of pain or ulceration.<sup>2</sup>

Carcinoma ex pleomorphic adenoma can be a challenging to diagnosis for the clinical features are similar to pleomorphic adenoma. Pathological assessment is the gold standard for making the final diagnosis that is based on the identification

of cell components of both pleomorphic adenoma and carcinoma under microscopy. The specimen can be obtained by fine needle aspiration or open biopsy. However false-negative finding may result due to sampling errors and only benign adenoma being identified.<sup>3</sup> The conclusion from image study is somehow elusive. Some articles suggested that those with malignant transformation have an irregular, infiltrative margin with or without associated malignant lymph nodes.<sup>3</sup>

Based on the histopathology, CXPA can be sub-classified into 3 subcategories: (a) non-invasive, (b) minimally invasive with less than 1.5 mm penetration of the malignant component into the extracapsular tissue, and (c) invasive with more than 1.5 mm of invasion from the tumor capsule to the adjacent tissues.<sup>4</sup> For our case, though an obvious capsule was found during operation, the histopathology showed the carcinomatous components has extended beyond the range of pleomorphic adenoma. So it's an invasive CXPA.

Table 1.

Article	Case No.	Site	Age	Male/ Female
Furukawa, et.al. <sup>6</sup> (2001)	1	Palate	51	female
Strick, et.al. <sup>7</sup> (2004)	2	N/A	N/A	N/A
Pires, et.al. <sup>8</sup> (2006)	2	N/A	N/A	N/A
Negahban, et.al. <sup>9</sup> (2006)	1	Palate	53	Male
Buchner, et.al. <sup>10</sup> (2007)	2	1 palate 1 floor of mouth	N/A	N/A
Li, et al. <sup>11</sup> (2008)	60	54 palate 6 floor of mouth	N/A	N/A
Chen, et.al. <sup>12</sup> (2010)	1	Palate	60	Male
Dyalram, et.al. <sup>13</sup> (2012)	1	Upper lip	72	Male
Sano, et.al. <sup>14</sup> (2012)	1	Buccal mucosa	71	Female
Kini, et.al. <sup>15</sup> (2012)	1	Buccal mucosa	17	Female
Gupta, et.al. <sup>16</sup> (2013)	1	Maxillary sinus	39	Female
Mitate, et.al. <sup>4</sup> (2013)	1	Upper lip	55	Male
Hong, et.al. <sup>3</sup> (2013)	1	palate	70	Male
Mariano, et.al. <sup>17</sup> (2013)	4	2 palate 1 upper lip 1 nasal cavity	N/A	N/A
Yamada, et.al. <sup>18</sup> (2013)	1	tongue	64	male
Nakamura, et.al. <sup>2</sup> (2013)	1	Buccal mucosa	52	Female
Sedassari, et.al. <sup>19</sup> (2014)	1	palate	74	Female
Bourell, et.al. <sup>20</sup> (2015)	1	palate	58	Male
Sum	83	63 palate 3 upper lip 3 buccal mucosa 7 floor of mouth 1 maxillary sinus 1 tongue	Average: 56.6	7/6

Note. N/A: not available.

The prognostic factors including T stage, lymph node involvement, histological grade, perineural invasion, extent of invasion,<sup>5</sup> and the status of surgical margin. The treatment of CXPA is usually through surgery with or without radiation therapy. Some author described that the rate of local recurrence for the submandibular and minor salivary gland CXPA is around 42% that is twice of that in the parotid gland.<sup>2</sup>

In current case, the clinical diagnosis by pre-surgical was pleomorphic adenoma. However, the surface ulcer and rapidly growth still implicated a risk of malignant tumor.

From the literature, the indication for postoperative radiation therapy depends on the tumor grade and the adequacy of the surgical margins. For this patient has a positive deep margin, postoperative radiation therapy was

strongly suggested. However this treatment plan was not accepted by the patient. After closely followed up for about one year, no local recurrence or metastasis was found. More long-term follow-up should be given for this patient.

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# 右側硬腭之多型性腺瘤中癌—病例報告

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## 摘 要

多型性腺瘤中癌是一種罕見，具侵犯性，尚未充分了解的一種惡性唾液腺疾病，特別是在小唾液腺中更少。絕大多數的病例都發生於大唾液腺中，最好發的部位是腮腺，其次為下顎下腺，小唾液腺則非常罕見。多型性腺瘤中癌的臨床表徵跟多型性腺瘤很相近，所以術前很難做出正確的診斷，必須要經由病理切片的判斷才能診斷，所以病理切片是為此疾病最標準的診斷工具。多型性腺瘤中癌的治療，目前仍以手術搭配放射線治療為主。

本病例為一個45歲之男性，自述於發現右側硬 有一無痛腫脹數個月，並且在腫脹的中央處有潰瘍的發生，經由口內切除此病灶，並於腫瘤邊緣並留有五公厘的安全距離。術後追蹤一年至今，在門診追蹤並沒有腫瘤復發的狀況。

**關鍵詞：**多型性腺瘤中癌，小唾液腺。

Received: September 08, 2015

Accepted: November 23, 2015

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