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Research Article

A RARE MALIGNANCY OF MINOR SALIVARY GLAND ON HARD PALATE- AN UNSUAL CASE REPORT

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ABSTRACT

Carcinoma Ex Pleomorphic Adenoma (CXPA) is a rare malignancy of salivary glands. CXPA arising in minor salivary glands is extremely rare. CXPA is very difficult to identify before surgical excision except for pathologic examination. It is because the clinical features of many cases are similar to those of pleomorphic adenoma (PA). Pathological examination is still the gold standard for its diagnosis. Surgical ablation combined radiation therapy are still the major treatment modalities of CXPA. The purpose of this article is to present a rare case report of carcinoma ex pleomorphic adenoma on minor salivary glands.

Key words:- Carcinoma ex pleomorphic adenoma, Hard palate, Minor salivary gland.

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INTRODUCTION

Carcinoma ex pleomorphic adenoma is a rare, aggressive, poorly understood malignancy that usually develops in a primary or recurrent pleomorphic adenoma[1]. CXPA contains elements of a benign pleomorphic adenoma and a frankly malignant epithelial component [2]. It accounts only 3.6% of all salivary gland tumors and 11.6% of all malignant neoplasms of salivary gland[3]. It has been reported that the most frequently affected site is the parotid gland, while the incidence rates in minor salivary gland are much lower[4]. 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 have reported a case of carcinoma ex pleomorphic adenoma in hard palate.

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Case report

A 50 years old male visited to our hospital with a chief complaint of oro-nasal communication since 3 years and also presented with growth in the hard palate since 6 months. A previous history of large growth in the same region 3 years back which he underwent surgery and postoperatively an obturator was placed which he discontinued on a later date. Later he developed with functional disturbance of slurred speech. Patient has reported to our hospital for further review. On extraoral examination there is a surgical scar in the right side midfaceregion resulting in a pseudo-exophthalmoses of right eye.

On inspection there is an oro-antral communication of the hard palate region in the right side measuring approximately 5x4cm in size. There is also a

single, large ,diffuse ulcerative growth present distal to the oro-antral communication of the hard palate measuring 2x2cm in size approximately extending medially from the distal aspect of the perforation to 3 cm away from the left alveolus, anteriorly it extends from 4 cm behind the rugae to 3cm infront of soft palate. The surface is grayish-black and the surrounding area is erythematous. The lateral wall of the oro-antral communication shows brownish discoloration. On palpation, the surface is rough, nontender on palpation. The growth is soft in consistency on palpation.(Figure 1).

Based on the history and the clinical findings the case was provisionally diagnosed as residual tumor in right side of hard palate. Incisional biopsy done on the ulcerated growth with local anesthesia (Figure 2). The specimen was sent to histopathological evaluation which showed hyperparakeratinised stratified squamous epithelium with fibrovascular connective tissue. The lesional connective tissue shows aggregates of plasmacytoid and epithelial cells in background of thick irregularly arranged collagen fibres. The cells are of varying size and shape exhibiting nuclear hyperchromatism and nuclear atypia, focal areas of necrosis and epithelial cells exhibit loss of cohesiveness suggestive of carcinoma ex pleomorphic adenoma (Figure 3 and Figure 4).

The patient was further evaluated for radiological examination in which computed tomography (CT) was taken. CT of axial section reveals a large expansile hypodense mass seen extending from left side of hard palate to right side which crosses the midline suggestive of a benign neoplastic pathology(Figure 5). Surgical management of total exicision of the lesion with left hemimaxillectomy done under general anesthesia(Figure 6). Excisional biopsy of the given H & E stained soft tissue shows orthokeratinised stratified squamous epithelium with fibrovascular connective tissue. The lesional connective tissue shows aggregates of plasmacytoid and epithelial cells in background of thick irregularly arranged collagen fibres. The cells are of varying size and shape exhibiting nuclear hyperchromatism and nuclear atypia suggestive of malignancy. The epithelial cells exhibit loss of cohesiveness. Focal areas of clear cells are seen suggestive of carcinoma ex pleomorphic adenoma (Figure 7). Patient reviewed after 6 months (Figure 8).

Fig 1. Intraoral view (mirror view)

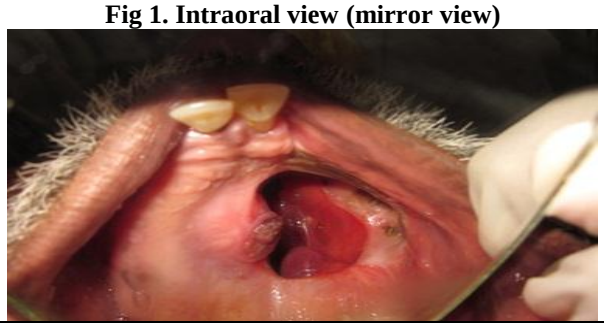

Fig 2. Incisional Biopsy specimen

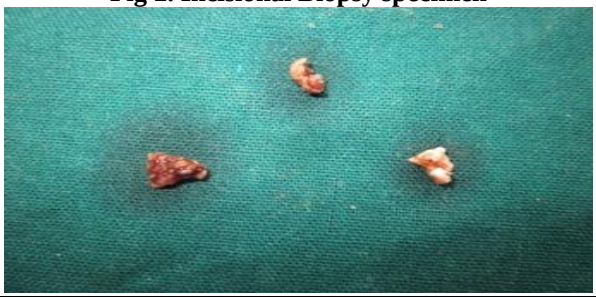

Fig 3. Histopathology(10X)

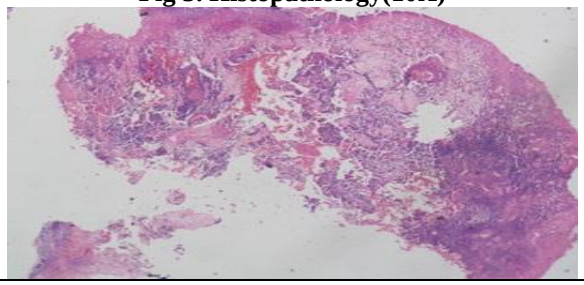

Fig 4. Histopathology (40X)

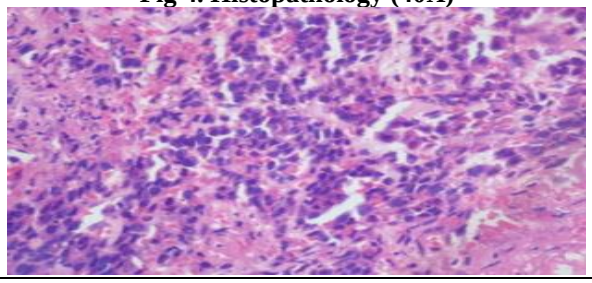

Fig 5. CT

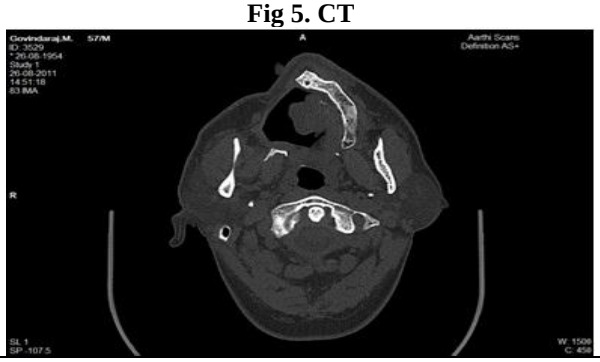

Fig 6. Excisional biopsy specimen

Fig 7. Histopathology of excisional biopsy specimen(40X)

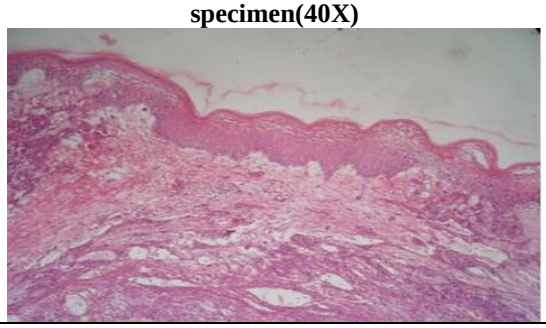

Fig 8. Post operative view

DISCUSSION

Carcinoma ex pleomorphic adenoma (CXPA) is a rare malignancy of head and neck region, in particular, for those in the minor salivary glands. This type of tumor usually arises in major salivary glands, most commonly in parotid gland, followed by submandibular gland. CXPA arising in minor salivary glands is extremely rare. From 2000 till now, only 83 cases have been reported. 63 of them occurred at palate. Carcinoma ex pleomorphic adenoma (CXPA) is defined as a carcinoma derived from preexisted pleomorphic adenoma[5]. In case of minor salivary glands (20%) palate is commonly affected[6]. The pathogenesis has not been well understood [6, 7].

Carcinoma ex pleomorphic adenoma been named as carcinoma ex mixed tumour, carcinoma ex adenoma, and carcinoma ex benign pleomorphic adenoma. It is found predominantly in the sixth to eighth decades of life and is slightly more common in females. In addition to these sites, cases of CXPA has been reported in the breast, lacrimal gland, trachea, and nasal cavity. Based on the presence and extent of invasion of the carcinomatous component outside the fibrous capsule, CXPA can be subdivided into non-invasive CXPA, minimally invasive CXPA, and invasive CXPA. CXPA can also be divided into those with only epithelial (luminal) malignancy and those with myoepithelial (non-luminal) malignancy. According to WHO histological classification (2005), the malignant mixed tumors, should be divided into 3 different clinical and histological entities: 1) carcinoma ex pleomorphic adenoma, 2) Carcinosarcoma, and 3) metastasizing pleomorphic adenoma. The term 'Malignant mixed tumors' is used in synonymous to CXPA, as the majority of them are composed of CXPA only. Clinically, it usually presents as a painless mass that is very similar to pleomorphic adenomas(PA). CXPA is very difficult to identify before surgical excision for pathologic examination is still the gold standard for diagnosis. Currently, surgery and post-operative adjuvant radiation therapy are accepted to improve local tumour control and increased survival. 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, regional and distant metastases are predictive of extremely poor prognosis. 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 [6-8].

The treatment of CXPA is usually thorough 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. From the literature, the indication for postoperative radiation therapy depends on the tumor grade and the adequacy of the surgical margins. More longterm follow-up should be given for these patients [5-8].

When CXPA involves the facial nerve, the patient presents with facial nerve paresis or palsy. The malignant component of CXPA is most often adenocarcinoma not otherwise specified. Sometimes, the component may be adenoid cystic carcinoma, mucoepidermoid carcinoma, or salivary duct carcinoma. The other less common histological subtypes include acinic cell carcinoma, epithelial-myoepithelial carcinoma, basal cell carcinoma, myoepithelial carcinoma, squamous cell carcinoma and clear cell carcinoma. The malignant component may also be a mixture of subtypes. A CXPA can be mistaken for a benign pleomorphic adenoma. It can also be misdiagnosed as other benign and malignant salivary gland tumours. A high grade salivary gland adenocarcinoma that is difficult to classify should include CXPA in its differential diagnosis. Metastatic mixed tumour is less common than CXPA and is differentiated from CXPA by having no carcinomatous component. Treatment for CXPA often involves an ablative surgical procedure which may or may not be followed by reconstructive surgery [8].

Malignant changes in PA have been associated with long duration, tumor size, tumor recurrence, radiation therapy and advanced age. Pain may be due to penetration of tumor into surrounding tissues. Other symptoms include facial nerve palsy, skin ulceration and fixation, lymphadenopathy, and dysphasia. Very rarely, soft palate CXPA presents as direct cavernous sinus (CS) invasion [6-9].

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Nil

CONFLICT OF INTEREST

No interest

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