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CASE REPORT

Pleomorphic Adenoma of Soft Palate: Unusual Occurrence of the Major Tumor in Minor Salivary Gland—A Case Report and Literature Review

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Abstract Salivary gland tumours constitute about less than 4 % of all head and neck tumours. Pleomorphic adenoma, also called benign mixed tumour, is the most common tumour of the salivary glands. About 80–90 % of these tumours occur in the major salivary glands mainly parotid gland and 10 % of them occur in the minor salivary glands. The most common site for pleomorphic adenoma of the minor salivary glands is the palate, followed by the lips and the cheeks. Other rare sites include the floor of the mouth, tongue, tonsil, pharynx, retromolar area and the nasal cavity. Here, we are reporting a case of pleomorphic adenoma of the minor salivary glands of the soft palate in a 36-year-old Indian female. The mass was removed by wide local excision with adequate margins under general anesthesia. There was no recurrence seen after a follow-up period of 1 year.

Keywords Pleomorphic adenoma (PA) · Metastasis · Fine needle aspiration cytology (FNAC) · Mixed tumor

Introduction

Pleomorphic adenoma (PA) is the most common neoplasm of the large salivary glands and affects mostly the parotid gland, less frequently the accessory salivary glands. It derives its name from the architectural pleomorphism seen under light microscopy. It is also known as "mixed tumor, salivary gland type", which describes its pleomorphic appearance as opposed to its dual origin from epithelial and myoepithelial elements. Mixed tumor accounts for 73 % of all salivary gland tumors. Corresponding to small glands, palate is the most common site for mixed tumor. Another region that is frequently affected by this tumor is the lips. A small minority of tumors are also located in the oral cavity, neck and nasal cavity [1–3]. Other intraoral sites include the buccal mucosa, tongue, floor of mouth, tonsil, pharynx, retromolar area, gingiva and nasal cavity [1, 4].

Pleomorphic adenomas may occur at any age, but mainly affect patients in the 4th to 6th decade. Male:female affliction ratio is 2:3 [5]. It also ranks as the most common salivary gland neoplasm in children, representing 66–90 % of all salivary gland tumors [6]. Wide local excision with removal of periosteum and involved bone is the treatment of choice.

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Case Report

A 36-year-old female patient reported to the Department of Oral and Maxillofacial Surgery, King George's Medical University, Lucknow, India with a slowly growing hard palatal mass on right side. The lesion frequently got traumatized while eating and had been present for the past two years. Clinical examination revealed non-ulcerated, dome-shaped, palatal swelling on her hard and soft palate junction involving faucial area and crossing the midline (Fig. 1). The mass was smooth, multinodular, firm, non tender and non fluctuant, on palpation it measured about 4 × 3 cm and did not involve any teeth CT scan was done to confirm the location, size and extent of lesion (Fig. 2). FNAC biopsy was performed, which was suggestive of PA.

Surgical Procedure

Patient was planned for surgery under general anesthesia. The patient was intubated through endotracheal tube via left nostril by fiber-optic guidance. Dingman retractor was applied and local anesthesia infiltered around the lesion. Mucosa around the lesion was marked and incised. Then a wide dissection was performed and the whole encapsulated tumor mass was excised along with the mucoperiosteum and overlying mucosa (Fig. 3). Surgical wound was primarily closed with advancement of adjacent mucosa, in layers in a water tight fashion (Fig. 4). There was slight shift of uvula to the right after final closure of wound but it did not affect the speech and swallowing of the patient postoperatively. Surgical splint was applied to reduce post operative palatal edema and hematoma. The patient's postoperative course was uneventful. No recurrence was observed after a follow-up of 1 year.

The excised mass was sent for histopathological examination, which revealed stratified squamous epithelium covering connective tissue. The underlying connective tissue showed cells arranged in sheets and duct-like pattern with a mucoid background confirmatory of PA (Figs. 5, 6).

Discussion

Salivary gland tumours constitute about less than 4 % of all head and neck tumours. These tumours are commonly seen in adults [7]. Among the various histological varieties of salivary gland tumours pleomorphic adenoma happens to be the commonest one constituting about 70 % of which 84 % occur in parotid, 8 % in the submandibular gland and 4–6 % in the minor salivary glands. 70 % of the tumors in minor salivary glands are PA, and the most common intra oral site is the palate, followed by the upper lip and buccal mucosa [8]. The unusual sites are sinuses, larynx, epiglottis, and trachea. PAs have also been reported in tongue [9], soft palate [10], vulva [11] and even external auditory canal [12]. The smaller the salivary gland that is affected, the more likely it is to trigger a malignant tumor. It most commonly occurs between fourth to sixth decades, however, reported instances in a 7-year-old patient and an 82-year-old patient are also seen [13]. Most common symptom manifested by tumors of this location are dyspnea, dysphagia, acute airway obstruction, and obstructive sleep apnea. However, in our case, tumor of size 5 × 4 cm

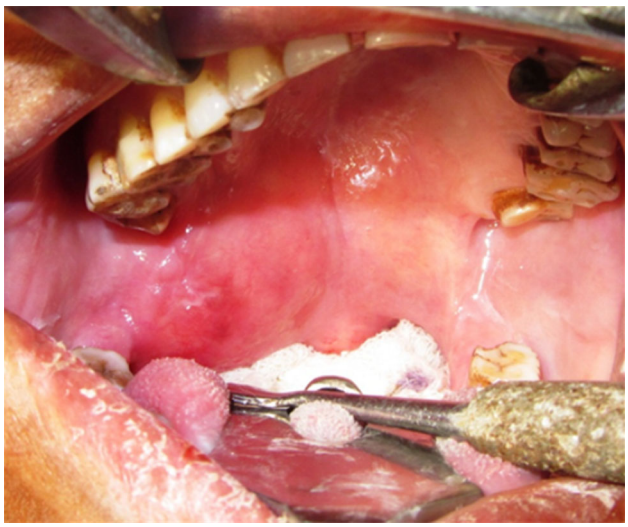

Fig. 1 Palatal swelling on hard and soft palate junction involving faucial area and crossing the midline

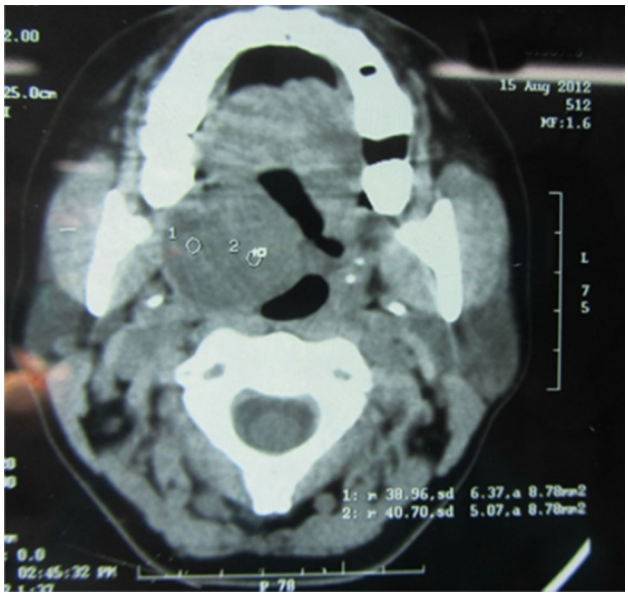

Fig. 2 CT scan of lesion showing extension of lesion

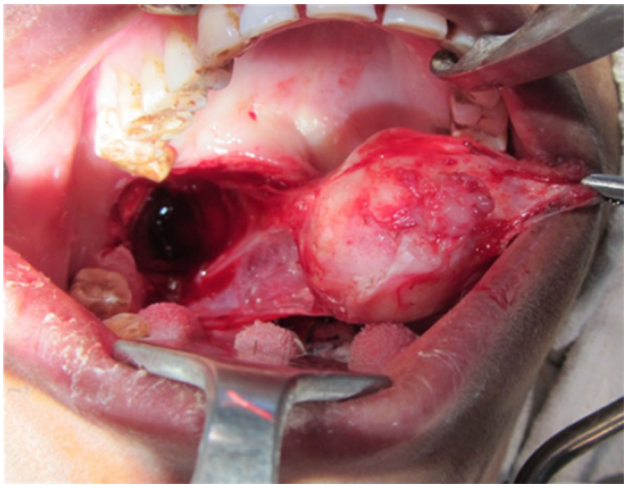

Fig. 3 Surgical exicision of the lesion

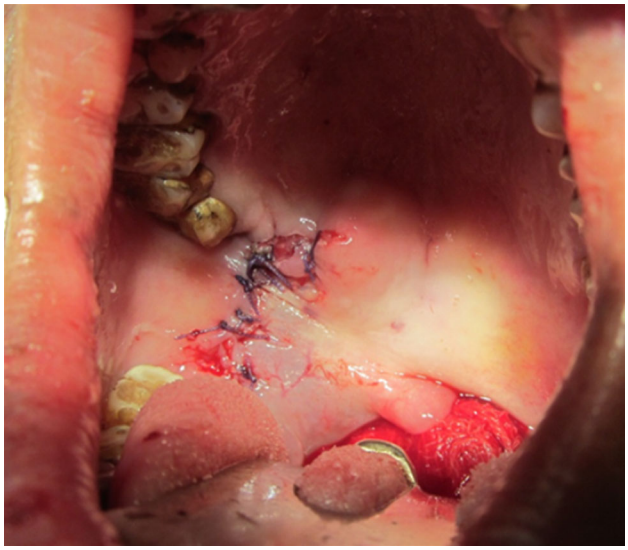

Fig. 4 Closure of surgical defect

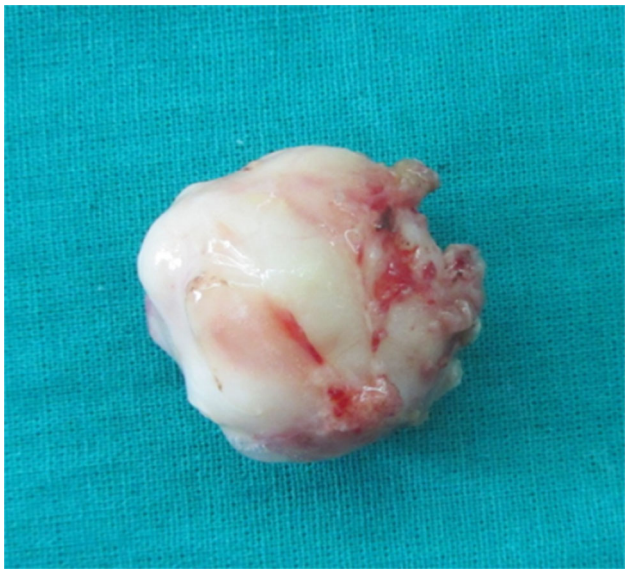

Fig. 5 Excised specimen

did not produce any respiratory distress although patient had history of slight dysphagia at times.

Various large sized PAs of hard palate have been reported in literature measuring about 7 × 6 cm [14], 5 × 4.5 cm [15], 3.2 × 4.2 cm [16]. Also, various large sized PAs from soft palate reported are 2.4 cm × 1.9 cm [10], 2.8 × 2.2 cm [17], 4.5 × 3 cm [18]. Our case measured PA of about 5 × 4 cm of soft palate; such a size of tumor is rare in this region. Thus, the unusual size and unusual location of this tumor, both parameters make this case a unique one.

Clinically, pleomorphic adenoma presents as a slow-growing, asymptomatic, unilateral firm mass that may become large if untreated. When originating in the minor salivary glands, in most cases, it occurs on the soft and hard palate due to the highest concentration of salivary glands here and is typically a firm or rubbery submucosal mass without ulceration or surrounding ulceration.

Anesthetic considerations during intubation and post operative airway management are important factors to be considered when planning surgery of tumors involving hard and soft palate.

Endotracheal intubation may be performed under deep inhalational anaesthesia or using muscle relaxants coupled with regional block. (Glycopyrrolate 0.1 mg intramuscularly and 2 drops of Xylometazoline into each of the nostrils can be given as premedication). Fibre-optic techniques can be used. A guide-wire may be threaded down the suction port of an adult endoscope, and the wire used to railroad a tube. Standard intravenous induction may be appropriate for older children or adults without anticipated airway difficulty e.g. propofol 4–6 mg/kg, thiopentone 3–5 mg/kg. A head ring and roll under the patient's shoulders extends the neck and tips the head down, and throat packs are used to absorb blood and secretions. Infiltration of local anaesthetic by the surgeon is recommended e.g. 1 % lidocaine with 1:200,000 adrenaline. This provides intraoperative analgesia, reduces blood loss and improves the surgical field. During surgery, a gag inserted over the endotracheal tube keeps the mouth open and tongue clear. Tube problems are common during surgery with a shared airway and may happen at any time. Vigilance is needed to prevent inadvertent extubation, intubation of the right main bronchus and tube kinking or occlusion. Controlled ventilation with muscle relaxation reduces anaesthetic requirements promoting a more rapid wake up and recovery of reflexes, as well as allowing lower PaCO2, which may reduce blood loss. Patient should be

extubated when fully awake while closely observing for signs of airway obstruction. Nasopharyngeal airways (NPA) can be effectively used and well tolerated in patients at high risk of postoperative airway complications, which may be inserted before emergence. The NPA can usually be removed the following day once swelling has subsided and the patient has mastered mouth breathing. Careful monitoring for 12–24 hrs should be done for early detection of any airway obstruction or postoperative bleeding.

Most important feature of pleomorphic adenoma of minor salivary glands is that there is no capsule, if present is only very thin [19]. This gives a false impression of infiltrating mass. These tumours are known to cause bone erosion i.e. are able to invade and erode adjacent bone, causing radiolucent mottling on the radiographs [20].

The potential risk of PA becoming malignant is about 6 % [6]. Pleomorphic adenomas involving minor salivary glands are painless [21] and slow in growth. Sometimes the growth rate could be fast. Rapid increase in size of the mass should lead to suspicion of intra lesional bleed/malignant transformation. The term pleomorphic adenoma is used to indicate the histological presence of both epithelial and mesenchymal tissues [22].

Histological features of pleomorphic adenoma include:

1. Islands of spindle cells over myxoid background.
2. Inner layer of epithelial cells.
3. Outer layer of myoepithelial cells

Three main histologic subgroups have been identified: myxoid (80 % stroma), cellular (myoepithelial predominant), and mixed (classic) type [23].

Histologically, it is highly variable in appearance. Classically, it is biphasic and is characterized by a mixture of polygonal epithelial and spindle-shaped myoepithelial elements in a variable background stroma that may be mucoid, myxoid, cartilaginous or hyaline. Epithelial elements may be arranged in duct-like structures, sheets, clumps or interlacing strands and consist of polygonal, spindle or stellate-shaped cells. Areas of squamous metaplasia and epithelial pearls may occur. The tumor is not enveloped, but is surrounded by a fibrous pseudocapsule of varying thickness [24, 25].

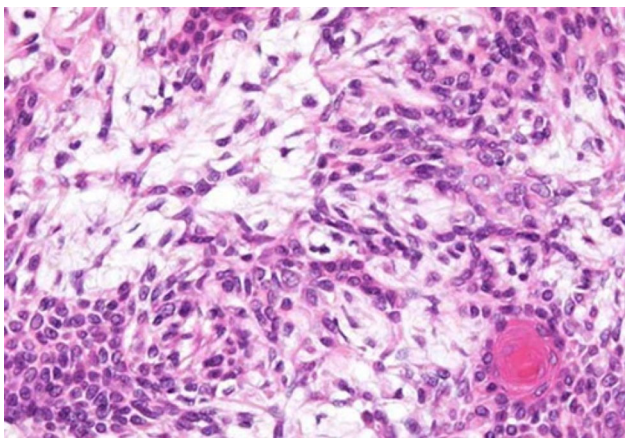

Fig. 6 Histological features include duct-like structures, sheets, clumps or interlacing strands and consist of polygonal, spindle or stellate-shaped cells

The main diagnostic modalities are FNA biopsy and imaging. Imaging helps in ruling out palatal erosion. Advanced diagnostic aids include USG, CT and MRI. Ultrasound is frequently used to guide FNA or core needle biopsy. CT is excellent for demonstrating bony invasion of palatal lesions. MRI provides superior soft tissue delineation/invasion and perineural spread. CT scan and MRI can provide information on the location, size and spread of tumor to surrounding superficial and deep structures and to some extent in determining type of tumor. In all radiological images of these lesions, one should look out for the presence of intact fat plane, the presence of which rules out malignancy [26].

The differential diagnoses for this case includes palatal abscess, odontogenic and non-odontogenic cysts, soft tissue tumors and salivary gland tumors. Palatal abscess was ruled out by clinical examination since the source of palatal abscess which is typically a non-vital tooth in vicinity or a localized periodontal defect was not found. In addition, this patient showed no signs of inflammation. Both odontogenic and non-odontogenic cysts were ruled out at the time of exploration since it did not demonstrate cystic nature. Palatal tissues contain components of soft tissue and harbour minor salivary gland tissues. As a result, soft tissue tumors such as fibroma, lipoma, neurofibroma, neurilemmoma as well as salivary gland tumors should also be considered in the differential diagnoses.

Research has shown epithelial origin of the mixed tumor, as well as clonal chromosome abnormalities with aberrations involving 8q12 and 12q15 [3]. The tumor often displays characteristic chromosomal translocations between chromosomes #3 and #8.

This causes the *PLAG* gene to be juxtaposed to the gene for β-catenin. Thus, this activates the catenin pathway and leads to inappropriate cell division.

The mainstay of treatment for salivary gland tumor is surgical resection of the tumor with a surrounding cuff of normal tissue. The excision should include periosteum or bone if these are included. If complete resection cannot be achieved, adjuvant radiotherapy should be added to improve local control. Generally, irradiation is reserved for inoperable cases [19]. These tumours are encapsulated and hence complete removal ensures cure. Care should be taken to leave at least 1 mm margins around the lesion. While removing the mass, rupture of the capsule is to be avoided to minimize recurrence.

The surgical approaches to the soft palate and parapharyngeal space include: Intra oral/transoral, transcervical, transparotid, transmandibular, and infratemporal. The choice of approach is to obtain adequate tumor visualization to ensure complete tumor removal with preservation of the surrounding nerves and vessels and to control any hemorrhage. Papadogeorgakis et al. [27] considered five points to be the main parameters in selecting the best approach while treating tumors of the soft palate and PPS.

- Proximity and the projection of the tumor to the oropharyngeal wall or the neck.
- Size of the tumor.
- Suspicion of malignancy.
- Vascularity.
- Relation of the tumor to the neck neurovascular bundle.

In our case, we preferred intraoral approach as it was a cosmetic approach and offers a direct access to the soft palate. Nowadays, this approach is reserved for well defined, avascular tumors <3 cm, tumors projecting in the oropharynx, and very near to the mucosa. Disadvantages of this approach are that it gives a limited exposure to the PPS with consequent unsatisfactory control of neck great vessels making it difficult in controlling massive hemorrhage, and increased incidence of nerve damage and capsular rupture leading to tumor recurrence [28].

Reconstruction of the palate is a challenging task. Soft tissue defects of the hard palate are essentially a nonissue, as the hard palate can be left to granulate. Bony defects in a dentate patient can be treated conservatively with an obturator. Bony defects of the upper alveolar ridge can cause a significant cosmetic and functional deformity, and therefore free tissue transfer techniques can augment the anterior projection of the face and soft tissue can be used to seal the oral cavity from the nose. Each technique has its advantages and disadvantages. The goals of maintaining speech, swallowing and anterior facial projection should be at the forefront of each surgeon's mind when approaching these difficult cases.

The "pedicle flap"—a technique for complete excision of benign salivary gland tumors of the palate—has been described in literature [29]. Immunohistochemical analytic study has been done to differentiate between polymorphous low-grade adenocarcinoma versus pleomorphic adenoma of minor salivary glands [30]. The management of salivary gland tumors of the palate has been documented by Pogrel [31]. Case of obstructive sleep apnoea has been reported secondary to PA of soft palate [32]. Myoepithelial cell predominant variety of PA of soft palate was described by Daryani et al. [10]. Various retrospective studies on PA have been documented in literature in Libyan population [33], Brazilian population [34], Iranian population [35] and Chinese population [36]. Diverse clinical, radiological, and histopathological presentation of pleomorphic adenoma was studied by Lingam et al. [37]. Detailed demographic study of intraoral minor salivary gland tumors was done by Waldron (1998) [38]. Another case of PA of soft palate was reported by Lomeo and Finneman [39].

Take Home Message are:

- Intraoral pleomorphic adenoma presents as a slowly growing painless mass, thus patients often seek late medical attention.
- Since the majority of minor salivary gland neoplasms arising in the palate are malignant, careful patient evaluation and preoperative diagnosis of pleomorphic adenoma by cytopathology and radioimaging is advised.
- Complete surgical excision with wide margin provides definitive diagnosis and treatment for this rare tumour.

Conclusion

To conclude, pleomorphic adenoma of minor salivary glands is relatively rare and therefore its diagnosis requires a high index of suspicion. Complete wide surgical excision is the treatment of choice. Early diagnosis and surgical excision results in complete cure with less or no morbidity compared to late diagnosis and surgery. Recurrence after many years of surgical excision as well as malignant transformation should be a concern and therefore long-term follow-up is necessary.

Conflict of interest None.

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