



Pleomorphic Adenoma of the Sublingual Salivary Gland - A Rare Occurrence

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Abstract

Sublingual salivary gland tumours are very rare and most, that occur, are malignant. A case of pleomorphic adenoma of the sublingual salivary gland, presenting as a slow growing swelling in the floor of the mouth, is presented for its rarity. The literature is also reviewed.

Keywords: Salivary gland tumour, Sublingual gland, Pleomorphic adenoma, Benign.

Introduction

Salivary gland tumours are rare, with an incidence of 2.5-3/1 lakh population, and those in the sublingual glands are rarer making up only 1% of all salivary gland tumors. A benign sublingual gland tumor is an even rarer entity as 70-90% of sublingual gland tumours have been reported to be malignant.^[1] Pleomorphic adenomas though the commonest salivary gland tumour, occurs mostly in the parotid gland (84%), 8% in the submandibular gland, 6.5% in the minor salivary glands and 0.5% in the sublingual glands.^[2] Till date there are very few cases of pleomorphic adenoma of the sublingual gland reported. Andreasen et al who studied sublingual gland tumours over 35 years in Denmark reported a total of only 29 cases, of which 28 were malignant and one was a benign pleomorphic adenoma.^[3]

Case report

A 31-year-old lady reported to our OPD, in March 2017, with a slightly painful swelling, that interfered with eating, in the floor of her mouth. The swelling was noticed 10 years ago, when it was smaller, and had been gradually increasing. She was on thyroid hormone supplements to correct her hypothyroidism and had no other positive medical, obstetric or surgical history. Intraoral examination revealed an oval swelling in right floor of mouth, 3.5 cm x 2 cm, extending into the submandibular triangle. It was firm, well circumscribed, not tender and non-pulsatile with few whitish petechiae on its surface. A patent Wharton's duct opening was seen medial to the swelling (Figure 1). Laboratory investigations were normal.



Figure 1 showing swelling in right side floor of mouth with patent Wharton's duct opening medial to swelling.

Ultrasonography (USG) showed a solid, well defined, focal mass in right submental space without calcification. Lesion was not infiltrating adjacent structure and separate from submandibular gland. Magnetic resonance imaging (MRI) showed a well-defined, lobulated lesion on right floor of mouth, measuring 4.4 cm × 2.5 cm × 4cm (AP × Transverse × Superoinferior). It extended upto the genioglossus muscle laterally, mylohyoid inferiorly and genioglossus medially. Laterally it abutted the lingual cortex of mandible without eroding it. The tumor was hypo to isointense on T1 and hyperintense on T2-weighted images with a heterogeneous post contrast enhancement. Ipsilateral dilated Wharton's duct with normal submandibular gland was seen. Fine needle aspiration cytology (FNAC) established the salivary gland origin of the tumour and further classified it as a pleomorphic adenoma (Figure 2).

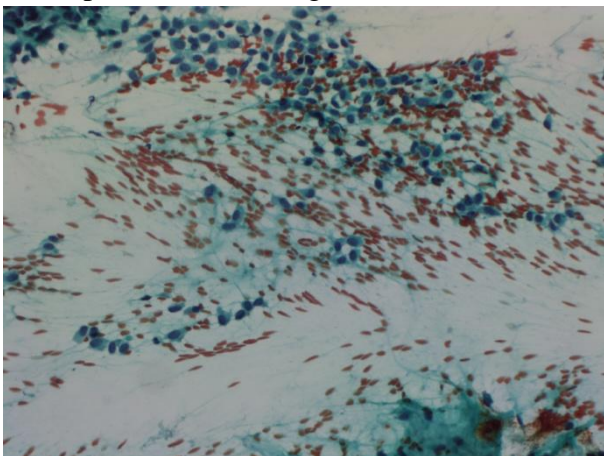


Figure 2: The epithelial cells are small round and uniform, some plasmacytoid with fine chromatin,

small nucleoli and delicate cytoplasm. The stromal tissue is seen in lower right. (PAPx200).

Right side sublingual gland mass excision was performed under general anesthesia. The mucous membrane of the floor of the mouth was incised anterior to Wharton's duct and flap raised, and sublingual gland and swelling resected as a single mass. The tumor did not adhere to the surrounding tissue (Figure 3).



Figure 3 shows lesion insitu.

Gross morphology was a well circumscribed and capsulated tissue 4 x 3 x 1.5 cm with external bosselation and congestion. On cut section grayish white to tan color firm, nodular tumor was seen with multiple fibrous septae in between giving a lobular appearance. At one end, adjacent 1x1x1cm soft area, resembling normal salivary gland tissue, was seen. (Figure 4)



Figure 4 Gross appearance shows a rubbery well circumscribed mass with a bosselated surface. Cut surface is solid with glistening areas indicative of

cartilaginous differentiation. Normal salivary gland is seen in the lower part of the specimen.

Histopathology confirmed the preoperative FNAC diagnosis of a pleomorphic adenoma. It was a well circumscribed encapsulated biphasic tumor composed of epithelial and mesenchymal components. The epithelial component was arranged as tubules, trabeculae, sheets of cells and acini having two cell layers. The mesenchymal component was composed of mucous, myxoid, fibromyxoid and chondroid tissue. The fibrous capsule showed occasional foci of tumor extension into the capsule. Mucinous salivary gland acini were seen adjacent to tumor capsule in one focus (Figure 5A-5C) Postoperative period was uneventful with good healing, a patent Wharton's duct and no neurologic damage.

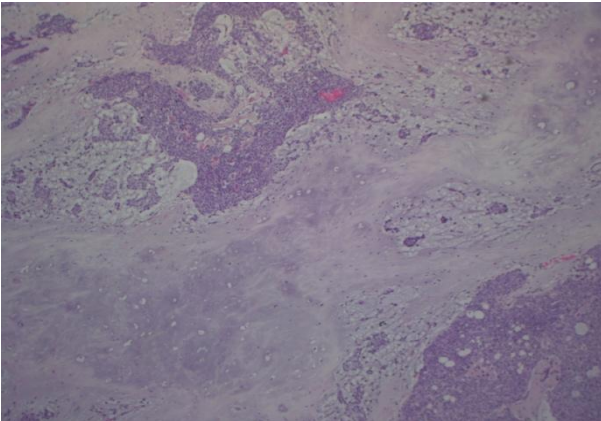


Figure 5A - Section shows the biphasic appearance of the tumour with admixture of the epithelium and stroma. Stroma is fibromyxoid with cartilaginous differentiation. (H&Ex40)

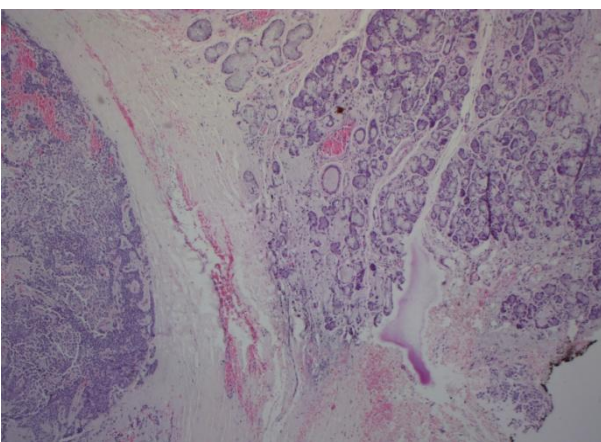


Figure 5B - Section shows the tumour margin with the normal predominantly mucinous salivary gland on the right. (H&Ex40)

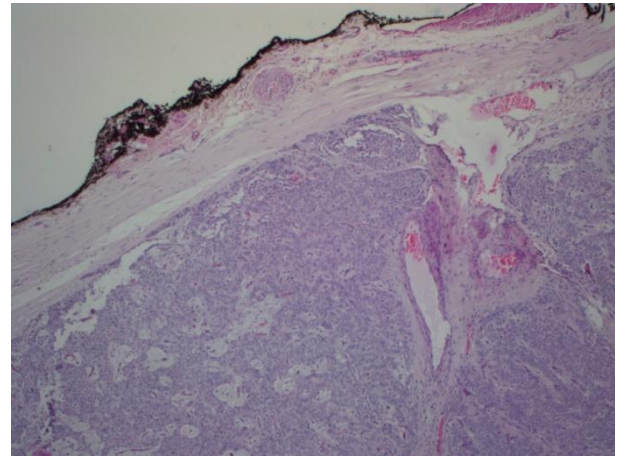


Figure 5C - Section shows capsule penetrated by tumour and extending till the inked surgical margin. (H&Ex40)

Discussion

The differential diagnosis in this case included non-neoplastic entities of salivary glands such as ranula and chronic sialadenitis, a dermoid cyst and neoplasms of salivary glands of submandibular/sublingual/minor salivary glands origin.

Exact preoperative evaluation of such a mass is necessary to diagnose, treat and prognosticate, but it can be challenging. USG has a high sensitivity (approximately 100 %) for differentiating intraglandular from extraglandular tissue.^[4] In our case, USG showed the tumour was solid, non-infiltrating and not involving the submandibular gland.

Fine needle aspiration cytology (FNAC) is a very useful method for evaluating suspicious salivary gland mass due to its low cost, minimum morbidity and rapid turnaround time. Meta-analysis of published series indicates 96% sensitivity and 98% specificity for neoplasia, whereas distinction between benign and malignant neoplasms has 79% sensitivity and 96% specificity^[5]. In our case FNAC could provide the diagnosis of a pleomorphic adenoma as typical constituents namely epithelial and myoepithelial cells in the characteristic fibrillary and chondromyxoid matrix were seen. In view of the relatively large size of the mass in this case sampling was done from different areas. The proportion of cellular constituents are extremely

variable from one Pleomorphic adenoma to another, and within an individual tumour hence it is important to sample different areas of the tumour with multiple passes^[6].

MRI is the investigation of choice to assess the exact extent, surrounding invasion, perineural spread and lymph node staging. It also eliminates dental artifacts and distinguishes tumour from obstructed secretions. It however cannot differentiate benign from malignant masses. Recently, new MRI techniques, such as dynamic contrast-enhanced MRI (DCE-MRI), diffusion-weighted MRI (DW-MRI), and proton MRI spectroscopy, have shown promising results in this aspect.^[7]

Histologically, a pleomorphic adenoma is also called benign mixed tumor as it consists of a biphasic population of epithelial and mesenchymal cells in varying proportions in a background stroma. Foci of microscopic extensions extending beyond the capsule may be seen.^[8] Hence wide local excision, and not enucleation, is the recommended treatment. Sun et al reported that for benign sublingual gland tumours, the resection of tumour and sublingual gland is the preferred treatment^[9]. The histopathology in our case showed occasional foci of tumour extension into the capsule and the patient has been kept on a regular follow up.

Although the etiology of pleomorphic adenoma is unknown, the incidence of this tumor has been found to increase 15-20 years after exposure to radiation.^[8] Pleomorphic adenomas are known to occur in all age groups and are more common in women. Overexpression of PLAG1, involved at 8q12 chromosome, is essential for the tumorigenesis of pleomorphic adenomas, although the mechanisms of over expression are variable.^[10] The prognosis for pleomorphic adenomas is good, but recurrence is reported in 0.4–5% of cases^[11] and malignant transformation occurs in about 6% cases.^[12] Soares et al have demonstrated that overexpression of mucin 1 gene (MUC1) was related to recurrence and malignant transformation.^[13]

Conclusion

Benign Pleomorphic adenoma of the sublingual salivary gland is extremely rare. Wide excision though recommended is difficult in case of large tumors due to location of the gland. Regular follow up is also recommended.

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