

Case Report

Pleomorphic Adenoma in unusual locations: Report of 4 cases

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Abstract:

Introduction: Pleomorphic Adenoma (PA) is a slow growing benign tumour of salivary glands, found most commonly in the parotid gland (80%), followed by submandibular gland (10%) and minor salivary glands (10%). In minor salivary glands, they occur mostly in the hard palate and soft palate followed by upper lip. Lacrimal gland PA is very rare. PA may be asymptomatic to incidental finding in some cases. Aetiology of PA is unknown but studies have shown that it has an epithelial origin and comprises cells from epithelial and mesenchymal differentiation. The standard treatment for PA is wide excision with adequate margins. It shows a recurrence rate of 2-44 % after excision.

Case Details: We report 4 cases of Pleomorphic Adenoma at unusual sites, that is, 1 case of lacrimal gland PA presented as right eye prominence and 1 case of recurrent swelling in the soft palate and 2 cases of slow growing swellings in upper lip. Histopathology of these lesions showed benign epithelial and mesenchymal components suggesting the diagnosis of Pleomorphic Adenoma.

Conclusion: Pleomorphic Adenomas are slowly progressive, benign lesions. Their occurrence in the minor salivary glands in unusual locations, should be kept in mind as these can be mistaken for other non neoplastic to neoplastic lesions.

Key Words: Pleomorphic adenoma, soft palate, upper lip, lacrimal gland

Introduction:

Pleomorphic Adenoma (PA) is a slow growing benign tumour of salivary glands, found most commonly in the parotid gland (80%) followed by submandibular gland (10%) and minor salivary glands (10%)^{1,2}. In minor salivary glands, they are seen mainly in the hard palate and soft palate followed by upper lip³. PA usually occurs in the fourth to sixth decade of life and are more common in women³. PA may be asymptomatic or painless, firm mass and in some cases can be an incidental finding too^{2,4}. The etiology of PA is not yet identified³. Epithelial origin and chromosome abnormalities with aberrations of 8q12 and 12q15 have been described in the literature³. Histological studies show PA to be comprising of cells with epithelial and mesenchymal differentiation⁴.

However, appearance of these benign growths may resemble the malignant tumors and may be misdiagnosed as malignant when PA shows increased cellularity and if there is any nuclear atypia in the myoepithelial component, or if the overlying mucosa shows pseudoepitheliomatous hyperplasia⁵. Proper distinction between benign and malignant growth is necessary from a prognosis and treatment perspective⁵. The recommended treatment for PA is wide local excision of the growth, the periosteum and involved bone⁵. A recurrence rate of 2 – 44 % after excision have been reported in studies⁵. The risk of the PA becoming malignant is about 1.5% when the duration is less than 5 years but the risk increases to 9.5% when the duration increases to more than 15 years⁶.

Case Report:

We report 4 cases of Pleomorphic Adenoma at unusual sites.

1. 53 year old woman presented with right eye prominence since one year, MRI scan suggested of a lacrimal gland tumour. Excised mass measured 2.8*2*0.8 cms. Histological features showed an encapsulated tumor with epithelial elements arranged in tubules, clusters, nests and trabecular pattern in a background of myxoid matrix. Foci of squamous metaplasia was also seen. Histopathological diagnosis of Pleomorphic Adenoma was done.

Fig 1. Pleomorphic Adenoma in lacrimal gland

2. 29 year old female who came with soft cystic swelling in *soft palate* measuring 4x3 cm. When she presented to us for the first time, she had come with a FNAC report of the same mass with a diagnosis of Pleomorphic Adenoma/ low grade adenoid cystic carcinoma. CT done showed a diagnosis of benign mixed tumor of salivary gland. Biopsy was done and histopathology report confirmed it as Cellular Pleomorphic Adenoma. Patient came for the excision of mass 9 months later and the MRI done at that time gave the diagnosis of benign mixed tumor of salivary gland.

Histopathology report of the excised mass was Cellular Pleomorphic Adenoma. Histological features were of a capsulated tumor where the epithelial elements showed increased cellularity and are composed of both round and spindle shaped tumor cells with fibromyxoid stroma.

Fig 2a. MRI scan showing swelling in the soft palate

Fig 2b. Cellular Pleomorphic Adenoma

3. 41 year old male with an *upper lip* swelling measuring 1.2x1 cm since 6 years
4. 43 year old female with a swelling in upper lip measuring 1.5x1 cm since 1 year (which was clinically diagnosed as retention cyst)

Excisional biopsy of both these cases (3 & 4) was confirmed as Pleomorphic Adenoma. squamous metaplasia was also noted in the last case.

Fig 3. Chondromyxoid area in Pleomorphic Adenoma

Fig 4. Pleomorphic Adenoma in upper lip showing minor salivary glands

Discussion:

Neoplasms of salivary gland constitute less than 3% of head and neck tumours³. Of all the salivary gland neoplasms, tumours of minor salivary gland comprise only 15-20%². Of all Minor salivary gland tumors, PA comprises 40-72%. However, de novo occurrence of benign PA of minor salivary glands is very rare². All of our cases showed Pleomorphic Adenoma arising in other sites without involvement of major salivary glands. PA of the minor salivary glands most commonly seen in palate, followed by lip, buccal mucosa, floor of mouth, tongue, tonsil, pharynx, retromolar area and nasal cavity³. Lacrimal gland PA is extremely rare condition⁷. Our cases showed PA occurring in lacrimal gland, soft palate and upper lip.

Aetiology of PA is not known, however studies mention that it is of epithelial origin³. Willis used the term 'Pleomorphic' for benign mixed tumor of salivary gland⁷. As the name indicates, histologically, PA shows a diversity of tissues where epithelial cells can be plasmacytoid in appearance, or arranged in a cord like cell pattern,

along with areas of squamous differentiation³. Myoepithelial cells are responsible for the production of abundant, extracellular matrix with chondroid, collagenous, mucoid, and osseous stroma³. PA associated with other features such as squamous metaplasia and keratin pearl formation could be mistaken for mucoepidermoid carcinoma and squamous cell carcinoma, both being malignant tumors⁸. In addition, Pleomorphic Adenoma and polymorphous low-grade adenocarcinoma are frequently mistaken with each other⁸. In one of our case (case no.2), patient had come with a fine needle report (outside) with the differential diagnosis of Pleomorphic Adenoma and low grade adenoid cystic carcinoma.

The risk of the PA becoming malignant is about 1.5% when the duration is less than 5 years but the risk increases to 9.5% when the duration increases to more than 15 years⁶. If PA is not treated, it can go for transformation into Carcinoma ex-pleomorphic adenoma and studies have shown that this is seen in 3% of minor salivary gland tumors². In case of lacrimal gland Pleomorphic Adenoma, other two epithelial tumors of lacrimal gland to be considered in the differential diagnosis are adenoid cystic carcinoma and adenocarcinoma⁷. Malignancy risk associated with Pleomorphic Adenomas of the lacrimal gland is 10–20%⁷. Studies have shown that 75% of Pleomorphic Adenomas transform into pleomorphic adenocarcinoma, and the remaining into cystic carcinoma⁷. Though, PAs of lacrimal gland are

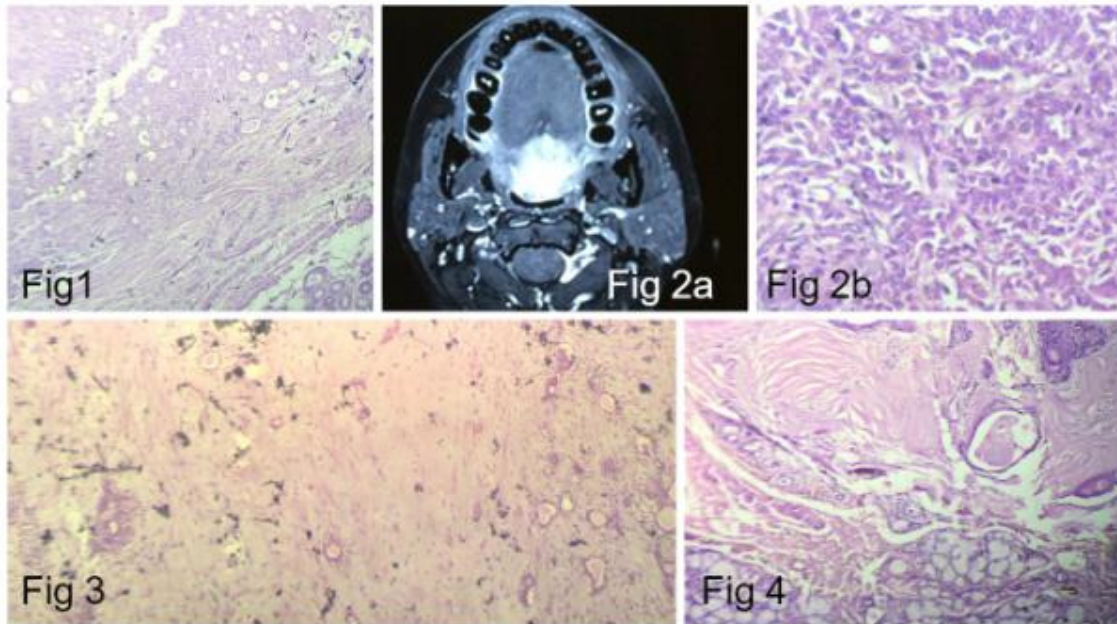
very rare and constitute only 4-9% all orbital tumors, when they occur morbidity associated with it is very high⁷. Factors which increase morbidity are when the tumor is displaced without its capsule and malignant transformations after incisional biopsies⁷.

CT scan and MRI help in locating the tumor, it's size and tumor extension to the adjoining areas, in particular to bone⁹. MRI can also help to facilitate differentiation of salivary gland tumors into benign and malignant⁶.

The recommended treatment for PA is wide local excision of the growth, the periosteum and involved bone⁵. PA is an encapsulated tumor with the capsule showing varying thickness and at times these tumors show lateral extension into the capsule^{2,3, 8}. Since Pa has a high risk of implantability, capsule rupture and incomplete excision would result in recurrence^{2, 3}. Studies have shown a recurrence rate of about 2-44%^{5, 8}. Recurrence is almost always as a result of incomplete excision with the tumor appearing in multiple foci and being more aggressive in nature, thereby requiring proper follow up^{2,5,8}.

Conclusion:

Pleomorphic Adenomas are slowly progressive, benign tumors but show wide diversity in morphological features which may challenge pathologists. Their occurrence in the minor salivary glands in unusual locations, should be kept in mind as these can be mistaken for other non-neoplastic to neoplastic lesions.



References

1. Jaber S, Rudic M, Keogh JJ. Pleomorphic Adenoma of the External Auditory Canal: A Rare Presentation. *Case reports in otolaryngology* 2015;696531.
2. Sood A, Chung S, Datiashvili RO. An Incidental Finding of Pleomorphic Adenoma of the Minor Salivary Glands in the Skin Area of the Lower Lip. *Eplasty*. 2014;14:317-321.
3. Debnath SC, Adhyapok AK. Pleomorphic adenoma (benign mixed tumour) of the minor salivary glands of the upper lip. *Journal of maxillofacial and oral surgery* 2010;9(2):205-208.
4. Verma P, Sachdeva SK, Verma KG, Sachdeva K. Pleomorphic adenoma of cheek: A rare case report and review of literature. *Indian Journal of Dental Research* 2014;25(1):122 -124.
5. Sengul I, Sengul D, Aribas D. Pleomorphic adenoma of the lower lip: A rare site of location. *North American journal of medical sciences* 2011;3(6):299-301.
6. Kato H, Kanematsu M, Mizuta K, Ito Y, Hirose Y. Carcinoma ex pleomorphic adenoma of the parotid gland: radiologic-pathologic correlation with MR imaging including diffusion-weighted imaging. *American Journal of Neuroradiology* 2008;29(5):865-7.
7. Binatli Ö, Yaman O, Özdemir N, Erdoğan IG. Pleomorphic adenoma of lacrimal gland. *Journal of surgical case reports* 2013;10:rjt089.
8. Jahangirnezhad M, Moghadam SA, Mokhtari S, Taravati S. Different histopathologic features of pleomorphic adenoma in salivary glands. *International Journal of Oral and Maxillofacial Pathology* 2013;4(2):7-11.
9. Khan MN, Raza SS, Zaidi SA, Hussain AK, Nadeem MD, Farid K. Pleomorphic adenoma of minor salivary glands. *Journal of Ayub Medical College Abbottabad* 2016;28(3):620-2.