



ORIGINAL RESEARCH PAPER

General Surgery

CASE REPORT OF A RARE PRESENTATION OF PLEOMORPHIC ADENOMA WITH SQUAMOUS METAPLASIA IN SUBMANDIBULAR SALIVARY GLAND: A DIAGNOSTIC CONUNDRUM.

KEY WORDS:

submandibular, salivary, tumor, pleomorphic adenoma, squamous metaplasia.

Dr. Anusha Lalithkumar*	Institute of General Surgery, MMC and RGGGH, Chennai- 600 003, Tamilnadu, India. *Corresponding Author
Dr. Shanthi Ponnandai Swaminathan	DGO., MS., Institute of General Surgery, MMC and RGGGH, Chennai- 600 003, Tamilnadu, India.
Dr. Vikas C. Kawarat	DNB., (General Surgery), Institute of General Surgery, MMC and RGGGH, Chennai- 600 003, Tamilnadu, India.
Dr. Kannan. R.	MS., Institute of General Surgery, MMC and RGGGH, Chennai 600 003, Tamilnadu, India.

ABSTRACT

Salivary gland tumors are quite uncommon in occurrence, and they constitute only about 1-5% of all the head and neck tumors. Submandibular salivary gland is affected in 5-10% of cases with the most common tumor being pleomorphic adenoma. A minority of cases may have squamous metaplasia with presence of keratin pearls, which may pose as a diagnostic challenge, especially while performing a fine needle aspiration study. A 29-year-old female came to our hospital with a 2-year history of a painless swelling in the left submandibular region. MRI neck showed a well-defined hetero intense lesion in left submandibular salivary gland. Fine needle aspiration revealed pleomorphic adenoma of left submandibular salivary gland. Final histopathology after excision confirmed the diagnosis of pleomorphic adenoma with squamous metaplasia, as an incidental finding. The most common diagnosis in a submandibular swelling is submandibular lymph node enlargement. Thorough clinical examination and evaluation is necessary to identify a submandibular salivary gland tumor, which is uncommon. Extensive squamous metaplasia in pleomorphic adenoma can be mistaken for malignancy, including mucoepidermoid carcinoma and squamous cell carcinoma. This can lead to diagnostic confusion and overtreatment. Very few studies have been conducted in submandibular salivary gland tumors, and further studies have to be carried out to know the nature of tumors affecting it.

INTRODUCTION:

Salivary gland tumors are quite uncommon in occurrence, and they constitute only about 1-5% of all the head and neck tumors¹. There are three pairs of major salivary glands- parotid gland, submandibular and sublingual glands, and multiple minor salivary glands. Majority of salivary gland tumors affect the parotid gland (70%). Out of that, pleomorphic adenoma constitutes 80-90% of the cases, making that the most common tumor affecting the parotid gland. Submandibular salivary gland is affected in 5-10% of cases with the most common tumor being pleomorphic adenoma^{2,4}. Classically, it is biphasic and is characterized by an admixture of polygonal epithelial and spindle-shaped myoepithelial elements in a variable background stroma that may be mucoid, myxoid, cartilaginous or hyaline. Pleomorphic adenoma has a pattern of cytogenetic abnormalities, most notably in the 12q13-15 chromosome region. PLAG 1 is the suspected gene, which has been localized to chromosome 8q 12. Being pleomorphic, it exhibits the ability to differentiate into epithelial (ductal and nonductal) cells and mesenchyme-like tissue (chondroid, myxoid and osseous)⁵. Mean age of occurrence is between the third and fifth decade of life⁶. The female to male ratio is 3.5:1, with increased female preponderance⁷. A minority of cases may have squamous metaplasia with presence of keratin pearls which may pose as a diagnostic challenge, especially while performing a fine needle aspiration study. Extensive squamous metaplasia in pleomorphic adenoma can be mistaken for malignancy, including mucoepidermoid carcinoma and squamous cell carcinoma. This can lead to diagnostic confusion and overtreatment.

Several studies have either focused on tumors of parotid gland or pleomorphic adenoma with squamous metaplasia in minor salivary glands, but only few literatures are available for submandibular salivary gland. Here, we present a case of a pleomorphic adenoma occurring in a submandibular salivary gland showing squamous metaplasia.

The work has been reported in line with the SCARE 2018 Criteria⁸.

Case Report:

A 29-year-old female came to our hospital with a 2-year history of a painless swelling in the left submandibular region. The swelling was insidious in onset, slow growing and painless. There was no history of fever or variation in size of the swelling in relation to meals. Past medical and personnel history was not contributory.

Extraoral examination revealed a swelling in the left submandibular region of size 3X2 cm, oval in shape, nontender to touch and firm in consistency, with well-defined borders (figure 1). Intraoral examination unremarkable, but the swelling was bimanually palpable. There was no evidence of reduced salivation from the left Wharton's duct. We arrived at a provisional diagnosis of a left submandibular salivary gland tumor based on the clinical findings.



Figure 1: Pre-operative image of a 29-year-old female showing left submandibular swelling- facial profile.

MRI neck axial and coronal sections showed a well-defined T2 hetero intense lesion in left submandibular salivary gland of size 3 x 2.1 x 2.4 cm (figure 2,3). Fine needle aspiration cytology revealed pleomorphic adenoma of left submandibular salivary gland.

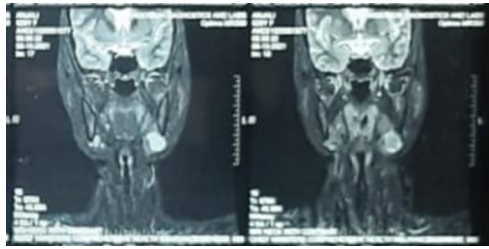


Figure 2: MRI Neck coronal view showing hetero-intense left submandibular gland mass- suggestive of pleomorphic adenoma of left submandibular gland.

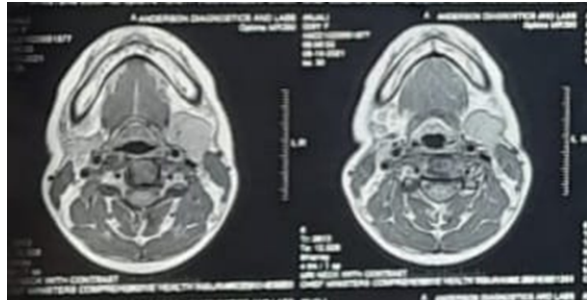


Figure 3: MRI Neck axial view showing hetero-intense left submandibular gland mass- suggestive of pleomorphic adenoma of left submandibular gland.

Patient was subjected to complete excision of left submandibular salivary gland and the specimen was sent for histopathological analysis. The microscopic picture showed sheets and cords of ductal epithelial cells, admixed with myoepithelial cells, in a chondromyxoid stroma (figure 4). Small islands of squamous metaplasia with keratin pearls was noted, but epithelium lined mucin-filled cysts were absent (figure 5). This ruled out the diagnosis of mucoepidermoid carcinoma. Squamous cell carcinoma was ruled out due to the presence of extensive morphological, epithelial and stromal diversity. Intracapsular/ in-situ malignancy of pleomorphic adenoma or carcinoma-ex-pleomorphic adenoma was ruled out primarily because, the tissue resected was in-toto, without any atypical changes or features of invasion. Secondly, there was absence of cellular or nuclear atypia, necrosis or capsular invasion. Final diagnosis of pleomorphic adenoma was given. Post-operative period was uneventful for the patient.

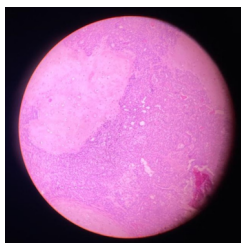


Figure 4: Histopathological image of the excised tumor mass showing dark stained tumor cells in chondro-myxoid stroma, in a predominantly mesenchymal background.

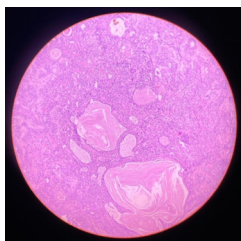


Figure 5: Histopathological image of the excised tumor showing islands of squamous metaplasia with interspersed keratin pearls.

DISCUSSION:

Pleomorphic adenoma is a mixed epithelial tumor with epithelial and mesenchymal background, with mucoid, myxoid, or chondroid tissue arranged in various configurations and embedded in a mucopolysaccharide stroma⁹. It is the most prevalent benign salivary gland tumor, accounting for 90% of all salivary gland tumors¹⁰. Submandibular salivary gland is the second most common site of pleomorphic adenoma¹¹.

The present case is a submandibular salivary gland pleomorphic adenoma, with squamous metaplasia. Several benign and malignant tumors show squamous metaplasia as an incidental histopathological finding. The cause is unknown, however it has been improperly linked to the intra-tumoral/tissue environment, such as trauma, infarction/ischemia, and infarction repair¹². The velocity and ease with which ischemia in the salivary glands causes a transition in the genetic programming of cytokeratin filaments could explain the varied degrees of squamous metaplasia, and hence ischemia appears to be the most likely origin for this alteration¹². Fine needle aspiration cytology for diagnostic purposes has been shown to induce the same in tumors during histopathological evaluation¹³. Fine needle aspiration was done in our patient, but it did not show any signs of ischemia and the aspirated tumor cells exhibited clear cut features of pleomorphic adenoma. Many-a-times pleomorphic adenoma with extensive or florid squamous metaplasia, when subjected to fine needle aspiration study, many show squamous cells that may cause a diagnostic dilemma.

Foci of squamous cells is a principle feature of pleomorphic adenoma, owing to the mixed cellularity of the tumor. But extensive or florid squamous metaplasia is quite uncommon and can be misdiagnosed as squamous cell carcinoma. Fine needle aspiration or incisional biopsies are the major culprits for this misdiagnosis due to limited and selective sampling. If the chondro-myxoid stromal pattern is absent, or comparatively less in the fields studied, the diagnosis becomes even more challenging. Thus, it is imperative to understand its occurrence and importance to avoid overtreating the patient.

Several cases have been reported with florid metaplasia in pleomorphic adenoma of minor salivary glands of the palate, pharyngeal wall. A case report of a 50yr old male, with a swelling in hard palate for only 1 week, was diagnosed as pleomorphic adenoma with extensive squamous metaplasia¹⁴. The unusual duration of presentation, with the site of presentation may nudge us towards a malignancy but it proved to be a benign condition. Immunohistochemical markers may aid in the final diagnosis.

Diagnosis of pleomorphic adenoma requires physical examination, imaging, cytology and histopathology. FNAC and incisional biopsy can help determine the proper management regimen, but must be thoroughly sampled to rule out any misdiagnosis. Other supportive investigations like computed tomography scanning and magnetic resonance imaging can provide information on the location and size of the tumor and its extension into surrounding superficial and deep structures.

CONCLUSION:

The most common diagnosis in a submandibular swelling is submandibular lymph node enlargement. Thorough clinical examination and evaluation is necessary to identify a submandibular salivary gland tumor, which is uncommon. The most common tumor in submandibular gland is pleomorphic adenoma, which is seen in our case. Extensive squamous metaplasia can be mistaken for malignancy, including mucoepidermoid carcinoma and squamous cell carcinoma. This can lead to diagnostic confusion and overtreatment. Very few studies have been conducted in submandibular salivary gland tumors, and further studies have to be carried out to know the nature of tumors affecting it.

Conflict Of Interest:

None.

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None.

Consent:

Obtained.

Ethical Approval:

Not required.

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