



DEEP SPACE MYSTERY: A RARE ENCOUNTER WITH PARAPHARYNGEAL MYOEPITHELIOMA

Dr. M. Veera Kumar

MS ENT., Professor and HOD, Department Of ENT, GGH, Kurnool Medical College, Kurnool, Andhra Pradesh.

Dr. K. Siva Govindan

Post Graduate, Department of ENT, GGH, Kurnool Medical College, Kurnool, Andhra Pradesh.

Dr. K. Mamatha Devi

Assistant Professor, Department of ENT, GGH, Kurnool Medical College, Kurnool, Andhra Pradesh.

ABSTRACT

Introduction: Parapharyngeal space tumours are rare, accounting for only 0.5% of all head and neck masses, with approximately 45% arising from salivary glands. Among these, myoeithelioma is an uncommon entity, representing just 1% of salivary gland tumours. **Methodology:** We report a case of a 72-year-old female from Kurnool who presented with hoarseness of voice for 3 months, along with recent onset dysphagia and an oral cavity swelling. Clinical examination revealed a diffuse bulge in the left peritonsillar region. Imaging (CECT and MRI) showed a well-defined, heterogeneously enhancing soft tissue lesion in the left parapharyngeal space, measuring 5×2.8 cm. The lesion was excised trans orally, and histopathological along with immunohistochemical analysis confirmed the diagnosis of myoeithelioma. **Conclusion:** Due to its rarity and deep-seated location, parapharyngeal myoeithelioma presents both diagnostic and therapeutic challenges.

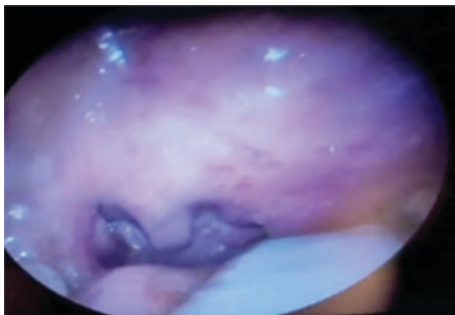
KEYWORDS : Myoeithelioma, Parapharyngeal Space Tumours**INTRODUCTION**

The parapharyngeal space is an intriguing compartment of head and neck which accounts for 0.5 % of all head and neck masses, attracts the special interest of surgeons, radiologists and pathologists due to the unusual pathology and proximity to vital anatomy. The majority of parapharyngeal space (PPS) tumours—approximately 70–80%—are benign, with 40–50% arising from the salivary glands, most commonly presenting as pleomorphic adenomas ⁽¹⁾. Tumours often originate from the deep lobe of the parotid gland and may extend into the PPS through the stylomandibular tunnel of Patey ⁽²⁾. Myoeithelioma represents about 1–1.5% of all glandular tumours. ⁽³⁾

In 1943, Sheldon introduced the term “myoeithelioma” to describe a group of tumours composed primarily of individual myoeithelial cells ⁽⁴⁾⁽⁵⁾. These are ectodermally derived cells situated between the basal surface of the acini, the basement membrane, and the surrounding intercalated ducts. They are also present in several other exocrine tissues, including the lacrimal glands, breast, prostate, and sweat glands ⁽⁶⁾. Within the head and neck region, they most frequently occur in the parotid gland (around 40%) and less commonly in the palate (about 2%). These tumours typically present as painless, slow-growing masses and are primarily diagnosed through histopathological examination rather than clinical evaluation.

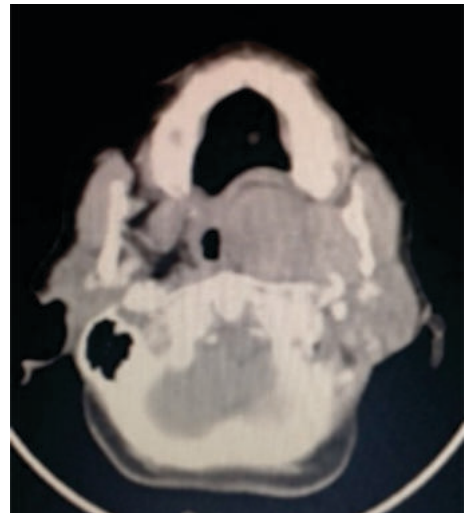
Case Report

A 72-year-old female from Kurnool district presented to ENT department with primary complaint of voice change persisting for the past three months. She also reported difficulty swallowing and the recent development of a swelling in the oral cavity over the previous week.

**Figure:1, Clinical Picture of Swelling.****Clinical Examination**

On examination, a diffuse swelling was observed in the left

peritonsillar region. The mass extended medially toward the midline, superiorly up to the soft palate, inferiorly down to the tonsillolingual sulcus, and anteriorly involved the anterior pillar. The swelling was firm and showed no plane of cleavage between it and the anterior pillar. Neck examination revealed no abnormalities.

**Figure:2, CECT Revealing the Mass.****Imaging**

Contrast-enhanced computed tomography (CECT) of the neck revealed a well-defined, heterogeneously enhancing soft tissue mass measuring approximately 5×2.8 cm located in the left parapharyngeal space. The imaging characteristics were suggestive of a neoplastic lesion.

**Figure:3, Resected Specimen**

Surgical Management

Under general anaesthesia, with the patient positioned in Rose's position, surgical excision of the lesion was performed through a transoral approach with endoscopic assistance. A 4 cm incision was made along the left anterior tonsillar pillar. The encapsulated cystic mass was carefully dissected from adjacent structures and excised completely. The postoperative period was uneventful, and the patient was managed with intravenous antibiotics. The excised specimen was sent for histopathological and immunohistochemical evaluation.

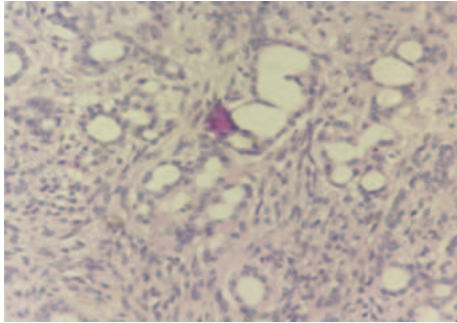


Figure:4, Histopathological Slide

Histopathological Findings

Microscopic examination showed glandular structures lined by cuboidal epithelial cells, along with focal areas of mucinous change. These features were indicative of myoepithelioma.

Immunohistochemistry

Immunoprofiling revealed tumour cell positivity for cytokeratin (CK), smooth muscle actin (SMA), vimentin, S100 protein, and p63, confirming the dual epithelial and myoepithelial differentiation characteristic of myoepitheliomas. The tumour cells were negative for TLE1, Desmin, Myogenin, CD117, and CD34. The Ki-67 proliferation index was less than 5%, supporting the diagnosis of a benign neoplasm.

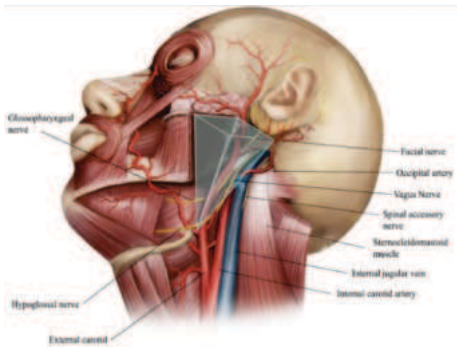


Figure:5, Parapharyngeal Space.

DISCUSSION

Parapharyngeal space is also known as pterygomaxillary space, pharyngomaxillary space, pterygopharyngeal space and lateral pharyngeal space. It is an inverted pyramid shaped potential neck space with apex lies inferiorly extending down to the greater cornua of the hyoid bone, base lies superiorly at the skull base, formed by the sphenoid and temporal bones and three surfaces (medial, lateral and posterior) and anterior leading edge in pterygomandibular raphe. Medially, it is bordered by the superior constrictor muscle and the buccopharyngeal fascia. Laterally, by medial pterygoid muscle, ramus of the mandible, deep lobe of the parotid gland, and the fascia of the posterior belly of the digastric muscle. Posteriorly limited by the prevertebral fascia, with the carotid sheath lying postero laterally and the retropharyngeal space postero medially. Two key fascial condensations of surgical relevance include the aponeurosis of Zuckerkandl and Testut, which extends from the styloid process to the tensor veli palatini muscle and divides the parapharyngeal space (PPS) into pre- and post-styloid compartments. The second is the stylomandibular ligament, stretching from the styloid process to the posterior border of the mandible, forming the stylomandibular tunnel of Patey and Thackray. This tunnel, located deep to the deep lobe of the parotid gland, provides a pathway for parotid tumours to extend into the PPS, often resulting in a dumbbell-shaped mass.

Parapharyngeal space (PPS) tumours are relatively rare and early reports of these rare tumours date back to 1963. The majority, about 45%, originate from salivary glands. Neurogenic tumours account for approximately 41%, followed by miscellaneous lesions making up 12%. Metastatic tumours are less common, comprising around 3%, with lymphoid lesions representing 2% of those cases. According to a study by Riffat et al., approximately 70 histological subtypes of parapharyngeal space (PPS) lesions have been identified, with 82% being benign and 18% malignant. The most common among these are salivary gland tumours, typically arising in the prestyloid compartment. Of these, the majority (75%) are benign, with pleomorphic adenoma comprising 64% of cases. These tumours often originate from the deep lobe of the parotid gland or from ectopic salivary tissue. Imaging, particularly CT or MRI, can help differentiate these origins—a fat plane between the mass and the parotid suggests a minor salivary gland tumour rather than one from the parotid's deep lobe. Over 15 histologic types of salivary gland tumours have been reported in the PPS. Deep lobe tumours are typically excised via a trans parotid and transcervical approach, while those arising from ectopic salivary tissue are usually removed through a transoral route, either endoscopic or robotic.

Myoepitheliomas are uncommon salivary gland tumours that originate from myoepithelial cells—specialized cells normally located between the epithelial cells and the basement membrane of salivary acini and ducts. Typically arise in the major salivary glands, occurrences in other regions of the head and neck are exceedingly rare. According to the World Health Organization (WHO) classification, benign myoepitheliomas are characterized by absence of chondroid or myxochondroid stroma. They have varied growth patterns depending on how the myoepithelial cells are arranged, which may include solid or reticular patterns, or a combination of both. In contrast, malignant myoepitheliomas are marked by significant cellular atypia and pleomorphism in the myoepithelial cells⁽⁷⁾. Clinically present as painless, slow-growing masses, on gross examination, they appear as solid, white or yellow, glistening masses. Myoepitheliomas of the minor salivary glands are generally non-encapsulated, whereas those arising in the parotid gland tend to be encapsulated.

Microscopically, myoepitheliomas are categorized into four histological subtypes: plasmacytoid, epithelioid, spindle, and clear cells, often seen in association with a mucoid or hyalinized stroma. The subtype, however, does not appear to impact the tumour's clinical or biological behaviour. The spindle cell variant is most found in the parotid, while the plasmacytoid type is more frequent in palatal lesions. To date, over 200 cases of benign myoepithelioma have been documented in English-language literature. According to the World Health Organization's classification, myoepitheliomas are considered to exhibit a more aggressive growth pattern compared to pleomorphic adenomas. Therefore, it's crucial to differentiate benign myoepitheliomas from malignant tumours such as malignant myoepithelioma, mucoepidermoid carcinoma, and spindle cell squamous carcinoma.

Definitive treatment involves surgical excision with a 0.5 cm negative margin. Bone involvement is rare but, if present, is managed similarly by ensuring a clear 0.5 cm margin. Ehrlich first introduced the transoral surgical approach in 1950, which is currently recommended only for small, nonvascular tumours due to its limited surgical exposure.⁽⁸⁾ However, in 1989, Goodwin and Chandler revived interest in this method by demonstrating its effectiveness, especially when combined with other approaches.⁽⁹⁾ They highlighted that it provides direct access to the parapharyngeal space (PPS) and allows better visualization and removal of deeply situated or larger tumours. In a similar context, we opted for the transoral approach in our case and found it to be a well-suited and effective method for management. The prognosis is excellent, and recurrence is rare after complete excision.⁽⁵⁾

CONCLUSION

With the advent of multiplanar radiological imaging including fine sliced CT with contrast and MRI with fat suppression and contrast, there is an improved preoperative diagnostic accuracy. This allows a better understanding of nature of these lesions and a better plan for surgical approach to them. Myoepithelioma is a rare presentation (1%) that provides both a diagnostic and therapeutic challenge because of its rarity and location. Accurate diagnosis and differentiation from malignant counterparts are essential for appropriate management. Surgical excision with clear margins remains the mainstay of treatment, with excellent prognosis and low recurrence rates when

completely removed. The transoral approach, though limited to select cases, can offer effective access for tumour excision in suitable patients.

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