CASE REPORT

Clear cell, giant myoepithelioma of tongue base: a diagnostic and surgical challenge

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SUMMARY

Clear cell variant is a rare histological type of myoepithelioma seen in parotid and soft palate. This article describes clear cell variant of myoepithelioma in the tongue base, which has not been reported in the literature so far. A 34-year-old man presented with dysphagia and foreign body sensation of throat. Video laryngostroboscopy using a 70° rigid telescope showed a smooth globular mass in the oropharynx arising from the tongue base. Based on clinical and radiological findings, the lesion was considered as benign. Fine needle aspiration cytology was not attempted fearing risk of bleeding, aspiration and airway compromise. Hence, an excisional biopsy followed by definitive histopathological examination without frozen section was planned. The patient underwent coblator-assisted excision and subsequently sent for histopathological analysis. There were cuboidal cell nests with abundant clear cytoplasm which stained positive for p63 by immunohistochemistry. This helped in establishing the diagnosis of clear cell myoepithelioma.

BACKGROUND

Myoepithelioma is uncommon and account for only 1% of all salivary gland tumours.¹ Initially thought as a variant of pleomorphic adenoma, myoepithelioma was identified as a separate entity in 1991 by the WHO.² The most common sites of origin include the parotid and the soft palate. However, other rare sites like the lips, inner lining of the cheek (buccal mucosa), floor of the mouth, gums and even the tongue have been reported in literature. They are usually slow growing and asymptomatic. However, very few cases of giant myoepitheliomas causing airway obstruction and dysphagia have been reported. Various histological variants of myoepithelioma have been described, some of which are spindle cell, plasmacytoid, epithelioid, clear cell and oncocytic, with spindle and plasmacytoid types were reported commonly.³ Clear cell variant of myoepithelioma is rare with only a few cases have been reported in the parotid and the soft palate. In this article, we have described a case of clear cell, giant myoepithelioma arising from the tongue base, which is reported in this subsite of the oropharynx for the first time in literature. We have also discussed the diagnostic and therapeutic challenges of a rare neoplasm that can present to the surgeon and the pathologist.

CASE PRESENTATION

A 34-year-old man presented to the outpatient department with irritation and foreign body

sensation in the throat for the past 6 months. He had dysphagia for solid foods for the past 1 month along with dyspnoea on exertion and on lying supine for the last 15 days. There was no history of voice change, throat pain, referred otalgia, fever or vomiting. He had no other comorbid conditions. On physical examination, his vitals were stable and he did not experience any respiratory distress. On examination of the oral cavity and oropharynx, a large globular mass was seen. The mass was occupying the entire oropharynx pushing the uvula anterior. However, a small area of posterior pharyngeal wall could be visualised on the left between the mass and the lateral oropharyngeal wall. Negotiating through this small area, video laryngostroboscopy using a 70° rigid Hopkin's rod telescope was carried out. A smooth globular mass was seen occupying the entire oropharynx causing narrowing of the oropharyngeal lumen with attachment to the tongue base (video 1). The laryngeal inlet and vocal cords were normal and mobile. The mass was firm in consistency and did not bleed on palpation. There was no palpable cervical lymphadenopathy.

INVESTIGATIONS

A contrast-enhanced CT scan of the neck showed a heterogenous, well-defined, 4×4 cm, lobulated, mildly enhancing lesion in the oropharynx. Inferiorly it extended up to the vallecula and crossing the midline with loss of fat plane over the pre-epiglottic space. Anteriorly, the mass extended up to the muscle plane of the tongue base without any signs of invasion (figure 1).

Fine needle aspiration cytology (FNAC) was not attempted because of the possible risk of airway compromise in case of accidental bleeding during the procedure. Hence, an excisional biopsy followed by

Video 1 Video showing a globular mass occupying the entire oropharynx causing narrowing of the lumen with attachment to tongue base.

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Figure 1 Contrast-enhanced CT scan with sagittal (A), axial (B) and coronal (C) views showing a heterogenous well-defined, lobulated, mild enhancing lesion in the oropharynx.

histopathological examination was planned. All baseline blood investigations, X-ray chest and ECG were normal. ELISA test for HIV virus was negative.

DIFFERENTIAL DIAGNOSIS

Considering the smooth appearance, slow growing and non-invasive nature of the tumour, the clinical diagnosis of a benign tumour was considered. The differential diagnoses included pleomorphic adenoma, basal cell adenoma, myoepithelioma, lipoma, neurofibroma, schwannoma and leiomyoma. Though the possibility of a malignant lesion like low-grade mucoepidermoid carcinoma, squamous cell carcinoma, lymphoma and slow-growing polymorphous adenocarcinoma was low, these tumours could not be ruled out completely.

Vascular tumours like haemangioma and arteriovenous malformations were ruled out based on the non-bleeding nature of the tumour and the mild contrast enhancement.

TREATMENT

Excisional biopsy followed by histopathological examination was planned. Endoscopic transoral coblation (controlled ablation) assisted excision of the tumour under general anaesthesia was planned. Consent for possible requirement for intraoperative or postoperative tracheostomy was obtained. Flexible nasopharyngoscopy guided nasotracheal intubation was done. Under general anaesthesia, Boyle Davis mouth gag with Doughty tongue blade was introduced and fixed. The tumour was visualised. To ensure complete removal, the tumour was resected in two parts. The oropharyngeal part of the tumour (extramucosal part) was visualised by 0° rigid Hopkins rod telescope and removed first using coblation. The second part which was attached and embedded submucosally in the tongue base could not be visualised by the 0° telescope. Hence, a 45° angled nasal telescope was used and the remnant of the tumour was removed completely. As the tumour was not infiltrating the surrounding tissues, it could be clearly distinguished and delineated from the tongue musculature and surrounding normal tissues by consistency and appearance. The tumour was removed completely and sent for histopathological examination with immunohistochemistry (IHC) for definitive diagnosis (figure 2).

OUTCOME AND FOLLOW-UP

Microscopic analysis of the tumour showed stratified squamous epithelium with the subepithelium showing a well-circumscribed lesion composed of nests and sheets of cuboidal cells, most of them with clear cytoplasm, intermingled with cells containing eosinophilic cytoplasm. Occasional hyaline globules were seen. There was no increased or atypical mitosis, nuclear pleomorphism and necrosis. These features were suggestive of a benign clear cell neoplasm (figure 3).

The benign clear cell tumours of the salivary gland include pleomorphic adenoma, myoepithelioma, oncocytoma, oncocytic hyperplasia and sebaceous adenoma. Immunohistochemical analysis was done for confirmation.

Figure 2 Completely excised mass specimen including both gross and submucosal parts.

Figure 3 Histological slide of specimen. (A) 40× magnification view shows a well-circumscribed lesion with a capsule. (B) 100× magnification view showing monotonous population, arranged in cords and small glands with clear cells visualised. (C) 200× power view showing many cells with clear cytoplasm with few showing eosinophilic cytoplasm. Nuclei are uniform, round to oval, vesicular, few showing nucleoli with no hyperchromatia, atypia or increased mitosis.

Immunohistochemistry

The tumour cells stained positive for p63 and were negative for vimentin and S100 (figure 4). Ki67 labelling index was 1%. These features were suggestive of a clear cell variant of myoepithelioma.

Follow-up

The patient was last seen 4 months ago. There was no evidence of residual or recurrent tumour. He was advised to review a year later, then once every 2 years if no signs of tumour recurrence.

DISCUSSION

Myoepitheliomas of the salivary gland were first described by Sheldon in 1943 and were initially classified as a type of pleomorphic adenoma.⁴ Myoepitheliomas are believed to arise from stem cells that differentiate into myoepithelial cells. Both benign and malignant types have been described in literature and the most common differential diagnosis is pleomorphic adenoma.⁵ Some authors believe that myoepitheliomas are a monomorphic, single cell variant of pleomorphic adenoma while others believe that they are a separate spectrum of benign tumours with monomorphic adenomas (composed of pure epithelial cells) at one end and myoepitheliomas (composed of only myoepithelial cells) at the other end.

Figure 4 Immunohistochemistry (IHC) markers. (A) Positive marker, P63. (B) Negative marker, vimentin and S100.

Despite FNAC being the next course of action, it was not done in this case fearing risk of fatal airway obstruction and aspiration.⁶ As clinical and radiological features were suggestive of benign neoplasm, the authors decided to proceed with an excisional biopsy followed by histopathological confirmation.

Benign tumours from oropharynx can arise from any subsite and the most common tumours are mucous retention cysts, papilloma and pleomorphic adenoma. Excision is the treatment of choice for these tumours. The authors had opted for excisional biopsy because of the lack of clinical and radiological evidence of invasion. The points suggesting benign pathology are chronic duration, well-defined margins, non-enhancing nature and absence of lymphatic spread. The confinement of tumour to tongue base without any signs of locoregional spread and invasion makes it a resectable tumour. Frozen section was not planned preoperative because the primary diagnosis was a benign tumour (pleomorphic adenoma) in this case.

Complete excision of any tongue base tumour is always a challenge because of the limited exposure, instrumentation and a high risk of bleeding. The excision becomes even more challenging in a setup without robotics. Bleeding from such sites can be decreased by using laser, radiofrequency or coblation.⁷ Coblation helps in volumetric removal of the lesion with minimal damage to the adjacent tissues thus reducing collateral damage.⁸ This feature makes it an ideal tool for surgeries in the upper airway including the oropharynx and the larynx.

Myoepitheliomas can appear in a wide range of people between the ages of 9 and 85 years, with no sex predilection. Currently, no risk factors have been identified. However mutations in a few genes like TP53 (seen in 25% of cases) and chromosomes 1, 9, 12 and 13 have been implicated. Myoepitheliomas commonly exhibit four phenotypes namely spindle cell (most common), epithelioid, plasmacytoid and clear cell (least common). Of these, plasmacytoid variant is more common in the subsites of the oral cavity.

Clear cell variants are extremely uncommon with only a few cases reported in the soft palate. They have polygonal cells with clear cytoplasm due to the high glycogen content. Sometimes, the clear cells can exhibit a signet ring-like or lipoblast-like appearance.⁹ On considering this diagnosis, it is important to rule out other clear cell neoplasms of the salivary gland and metastatic tumours with clear cells especially from the kidney and lung. Metaplastic changes and oncocytic differentiation may also be seen. All these features make the diagnosis of this variant extremely difficult.

That makes role of IHC quite significant in the diagnosis of these tumours. Myoepithelial cells may express S100, calponin, smooth muscle actin and p63 among other markers.^{10 11} As a nuclear marker, the presence of p63 is more sensitive. Positive staining for one or more of these markers along with the typical morphology will help in the diagnosis. In this case, the presence of cuboidal cell nests with abundant clear cytoplasm which showed strong positive nuclear staining for p63 by IHC helped us to arrive at a diagnosis of a clear cell variant of myoepithelioma.

CONCLUSION

Myoepithelioma arising from the base of the tongue is rare. Restricted access, airway complications and the requirement of expert surgeons and pathologists make the diagnosis and

treatment of this tumour difficult. This article describes a clear cell variant of myoepithelioma of the base of tongue, which has not been reported in literature so far.

Patient's perspective

I was informed by the doctors about my clinical condition. I have been informed after surgery that a rare neoplasm called myoepithelioma has been diagnosed in the back of my tongue. As my symptoms have resolved, i am quite happy about the outcome. I understand that reporting such a rare medical condition might be useful to the medical sciences and young upcoming doctors. I am happy that reporting my clinical condition in a journal will add to existing knowledge and help in the management of patients in future. Thank you.

Learning points

- Myoepitheliomas are rare neoplasms of tongue base.
- Restricted access and airway complications make the management of this tongue base tumour complicated.
- This article describes a clear cell variant of myoepithelioma in tongue base, which has not been reported in literature so far.

Contributors The first author, PKS is a Senior Consultant ,Professor and Resident guide in Sri Ramachandra University hospital, Chennai. He was the main surgeon of the patient in this study. He also helped the VM(Corresponding author) in the write-up of this article . His ideas and experience has helped in shaping up the article for possible publication. The second and corresponding author, VM is a senior resident and registrar in the same university hospital. He was the second main surgeon and did the main write-up of the article.He did an extensive research and reviews regarding myoepithelioma before beginning to write the article. The third author, RR is a third year resident trainee in Otolaryngology. She actively

involved herself in peri-operative management of this patient and also in collecting photographs of CT scan, histology slides and IHC slides. She also helped the corresponding author in preparing the main manuscript. Thank you.

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