Case Reports and Reviews

Case Report

Diagnostic Dilemma with Atypical Histological Outcome: A Rare Case of Paediatric Sublingual Gland Spindle Cell Myoepithelioma

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Abstract

In daily clinical practice, salivary gland conditions are not infrequently encountered by the ENT, oral, head and neck surgeons who often must work closely with the pathologists to manage them. Occasionally, an unusual histological diagnosis will be made which will demand a slightly different approach to management. More often encountered conditions include ranulas, epidermal/dermal inclusion cysts, lymphatic cysts, thyroglossal cysts, sialolithiasis and branchial cleft cysts all of which may present with floor of mouth swellings. There are a few rarely encountered histological patterns; here, we report a rare case of spindle cell myoepithelioma of the left sublingual gland in a child while reviewing literature to highlight its pattern of clinical presentation, diagnostic dilemma and management.

Introduction

Over decades, several histological diagnoses have been reported in the salivary glands. Most are benign, including floor of the mouth cysts - mucous retention phenomenon (mucocele, ranula), dermoid and epidermal inclusion cysts, thyroglossal duct cysts, branchial cleft cysts, hemangioma, lymphangioma, cystic hygroma as well as non-cystic conditions like sialolithiasis, lipoma and occasionally pleomorphic adenoma. Quite rarely, spindle cell myoepithelioma are encountered in the sublingual gland; we report one of these, experienced in our practice while reviewing available related literature.

Case Report

We report on the case of an otherwise healthy 8-year-old boy noticed to have a swelling in the left side of the floor of his mouth for about 6 months prior to visiting the hospital. Being of initial size of a peanut, it gradually grew to approximately the size of a cherry over the period. There was no associated pain or discharge from the swelling, however, there was difficulty maneuvering food bolus in his mouth.

Examination revealed a domed shape, pinkish, cloudy mass with mixed consistency occupying the entire left half of the floor of mouth, mobile and non-pulsatile. The tongue and the rest of the floor of the mouth and submandibular glands appeared normal. Needle aspiration yielded a dry tap in contrast to the typically thick, straw-coloured yield classical of ranula.

Subsequent ultrasound scan revealed a well circumscribed, solid mass with a smooth outline, hypoechoic texture and lobulated borders. The mass was excised intraorally with the dead space created irrigated with saline and closed in layers with vicryl 3/0 absorbable sutures.

Histopathological analysis of the excised mass was reported as macroscopically showing firm grayish white tissues, measuring 4 by 3cm by 2cm in its widest dimensions. Microscopic evaluation identified a mesenchymal neoplasm comprising interlaced fascicles of spindle-shaped, benign smooth muscle cells with plump, cigar-shaped nuclei and surrounding areas of eosinophilic cytoplasm. Also seen within the myxoid stroma were focal areas of lymphocytic infiltration and mitosis of <3 per 10HPF. There was notable absence of ductal or tubular elements. Overall features were in keeping with benign spindle cell tumor. There were also identifiable signs of chronic inflammation of leiomyomatous tissue. A histological diagnosis of spindle cell myoepithelioma was concluded.

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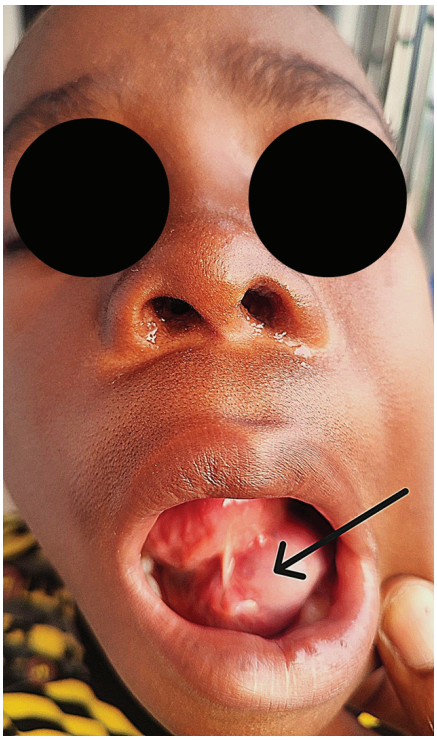

Figure 1. *Clinical photograph showing the mass in the floor of the mouth.*

Figure 3. *Gross pathology specimen of the excised mass.*

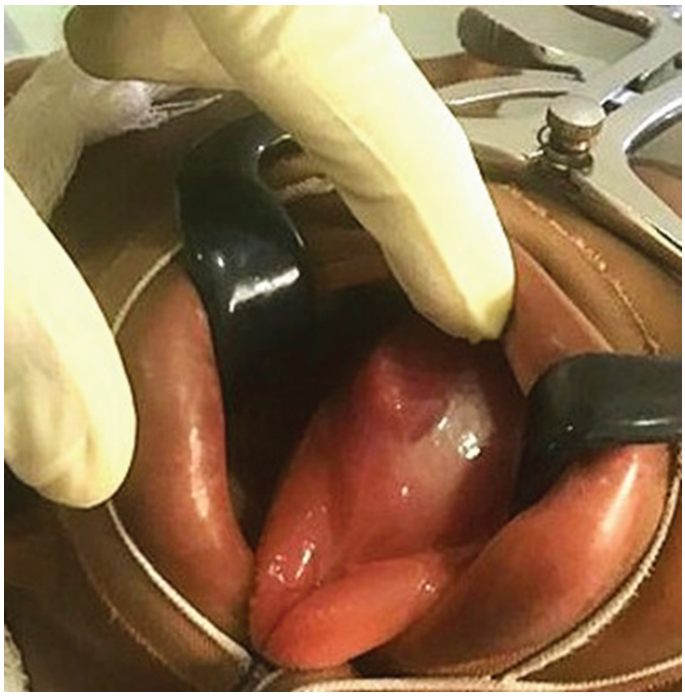

Figure 2*. Intraoperative exposure of the floor of mouth mass using Doyen mouth gag*

Discussion

The classification of myoepitheliomas is widely credited to Walter Sheldon in 1943 while the diagnostic criteria was defined first by Barnes et al. [1-3]. They account for approximately 1% of salivary gland tumours with about 40 % occurring in the parotid gland and 21% in minor salivary glands [3].

Myoepitheliomas have been described in various organs, notably in the oral cavity/oropharynx [4,5]. A spindled myoepithelioma of the sublingual gland is a relatively rare, benign occurrence and when present, encompasses almost entirely spindle shaped myoepithelial cells. They have also been reported, perhaps more commonly in parotid and submandibular glands and maybe to a lesser extent, in minor salivary glands. Overall, they are quite rare, accounting for less than 1% of all salivary gland tumours. They are mostly benign; however, malignant transformation can occur in some cases. There are also reports of malignant myoepitheliomas arising from pleomorphic adenomas of salivary glands [6-8].

The presentation of myoepitheliomas is often an insidious, slow-growing painless mass, in our case, to the left of the floor of the mouth. Majority of these patients are adults in the 20-50 years age range. We report our case due to the relative rarity of a pathology of this kind in a child. Over several months, the child began drooling with difficulty chewing and swallowing prompting hospital attendance. This is a familiar pattern as recorded in similar case reports. As reported widely in other works, the initial size of these lumps often ranges from around 1- 4cm and can slowly enlarge up to 10cm [3,9]. In our case, it was difficult to accurately estimate the size at onset given the age of the child, socio-cultural background and literacy level, peculiar to our setting. At presentation, the onset size was described as ‘about the size of a pea’ and on examination, it measured 4cm, domed shape, pinkish with mixed consistency and occupying the entire left half of the floor of mouth. It was non-tender, mobile and non-pulsatile.

Diagnosis of myoepitheliomas can be challenging due to its clinical similarity with other pathologies that can cause swelling in the floor of the mouth. In addition, the varied cell

morphologies histologically and the need to differentiate it from other spindle cell lesions can potentially pose diagnostic dilemma. Typical myoepithelioma appearances histologically comprise myoepithelial cells with varying growth patterns and appearances which may be solid, myxoid (in which case may mimic pleomorphic adenoma), reticular, or in fact, a mixture of these. Furthermore, appearances such as spindle-shaped, plasmacytoid, clear, and epithelioid have been described [10]. In addition, tumour cells may look like a few other conditions; example being spindle cell myoepitheliomas mimicking neural sheath tumors and clear cell variants sometimes mistaken for renal cell carcinomas or clear cell carcinomas. It is also often a challenge to differentiate a benign myoepithelioma from a myoepithelial carcinoma as Pathologists must rely heavily on spotting infiltrative behaviour, destructive growth patterns, necrosis and high mitotic index among others. For this reason, fine needle aspiration cytology (FNAC) is generally considered non-diagnostic when considered in isolation [11]. Making the correct histological diagnosis always requires astute histological assessment with the experience of specialized anatomic pathologists playing a key role.

The role of ultrasound in evaluating sublingual gland myoepithelial tumors is mainly for tumour localization, characterization, and biopsy guidance. The typical sonographic appearance is a solid, hypoechoic mass with heterogeneous internal structure often with well-circumscribed, lobulated, distinct borders, distinguishing it from its malignant counterpart which will have irregular or infiltrative margins [12]. The mainstay of diagnosis is histopathological assessment of the excised tumour.

The primary treatment of sublingual gland myoepithelioma is complete surgical excision best taken with a clear margin of healthy surrounding tissue. Surgical access in our case was intraorally, keeping the mouth gagged with Doyen's mouth gag. This procedure can be done under local anaesthesia but given the age of our patient, it was done under general anesthaesia. Following complete excision, the defect was closed in layers with absorbable sutures (vicryl 3/0). Management is similar in other reported works. The extent of resection generally depends on the suspected pathology or biopsy outcome. More radical procedures have been described as for malignant myoepitheliomas [13,14]. This can sometimes involve mandibulectomy with reconstruction with composite free flaps [13-15]. Our patient was discharged from hospital one day post operatively following overnight stay for observation. He was followed up 2 weeks post-operatively with good outcomes.

Conclusion

We report a relatively rare case of spindle cell myoepithelioma in an 8-year-old boy, with insidious onset and slow growth. It was managed by complete excision with review of the histopathological outcome. We have highlighted the rarity of this histological diagnosis in this age group and the associated diagnostic dilemma. Hopefully, we have underscored the need to keep clinical differential diagnosis wide enough to accommodate these kinds of rarities even in paediatric population and bearing in mind that multidisciplinary approach will always be the desired pathway of management for best outcomes.

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