RARELY ENCOUNTERED TUMORS OF THE MAXILLOFACIAL REGION – A CASE SERIES

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ABSTRACT

A variety of distinct lesions from differing tissues of origin are encountered in the Maxillofacial region. Though rare, the clinical behavior and management of entities such as Hidradenoma, Neurofibroma and Low grade Mucoepidermoid Carcinoma should be well understood, and it is important to consider them in differential diagnoses. In this report we present the clinical features, histopathology and surgical considerations in the management of these rare lesions.

Keywords:Hidradenoma, Neurofibroma, Mucoepidermoid carcinoma.

INTRODUCTION

A wide variety of non-odontogenic tumors, both benign and malignant, are encountered in the maxillofacial region. Whereas, oral squamous cell carcinoma is most commonly occurring malignant tumour [1] , tumors like Hidradenoma , intraoral neurofibroma, mucoepidermoid carcinoma of sublingual salivary gland are not seen very often. In this paper we compile the clinical presentation and management of these rarer entities.

Hidradenoma

Tumors arising from skin adnexa are encountered in the head and neck. 'Hidradenoma' arise from sweat glands. These lesions may also be known as 'solid-cystic hidradenoma', 'clear cell hidradenoma' or 'eccrine sweat gland adenoma'. [2] The characteristic features of Hidradenoma are its slow, gradual growth and a history of serous discharge. Ulcerated lesions may resemble basal cell carcinoma. [3] This is of clinical significance because when head and neck surgeons encounter this rare tumour, it may be taken for basal cell or squamous cell carcinoma. [4] Hence, it is important

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to elicit a detailed history and carry out a good clinical evaluation of the lesion. These lesions are best treated by surgical excision and recurrences are rare.

We present a case of hidradenoma which occurred on the left nasolabial fold.

Case Report

79-year-old female was referred for the evaluation of a swelling on the left nasolabial fold. The swelling had been asymptomatic, present for about 5 years, with growth since 1 week. Inspection revealed a warty swelling 2.5X1cm, pearly white with crusting and centrally filled with a clear fluid. Surrounding area was non-erythematous. On palpation, lesion was firm with a rough surface (Figure 1).

A provisional diagnosis of basal cell carcinoma was made. Other lesions kept in mind were papilloma, squamous cell carcinoma and keratoacanthoma.

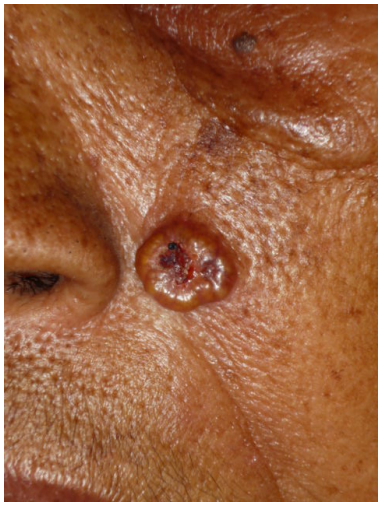

Figure 1. Lesion seen on left nasolabial fold

Under monitored anesthesia care, the lesion was excised with a minimum 5 mm safety margin all around. Primary closure was achieved after soft tissue advancement (Figure 2). Patient had an uneventful recovery and sutures were removed after 7 days.

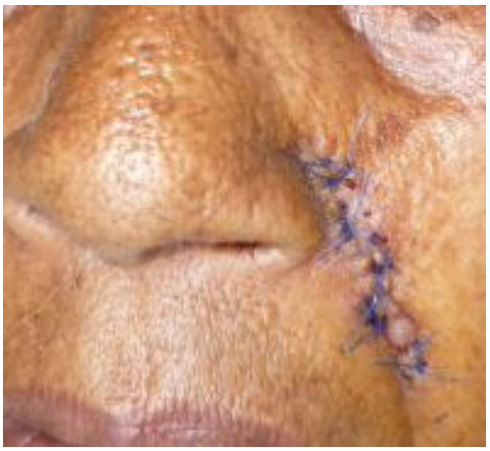

Figure 2. Primary closure after excision

Histopathology revealed an unencapsulated, well circumscribed lesion underlying atrophic orthokeratinized epithelium. Lobulated masses having biphasic architecture of round cells with a vesicular nucleus intermixed with clear cells having a smaller nucleus were seen (Figure 3). Few areas showed cystic degeneration. The tumour also had epidermal connections, with PAS positive material strewn between the cells (Figure 4). The connective tissue between the lobulated mass was sparse containing thin collagen fibers and an infiltration of inflammatory cells consisting of neutrophils, lymphocytes and mast cells.

These features were suggestive of a histopathologic diagnosis of Hidradenoma.

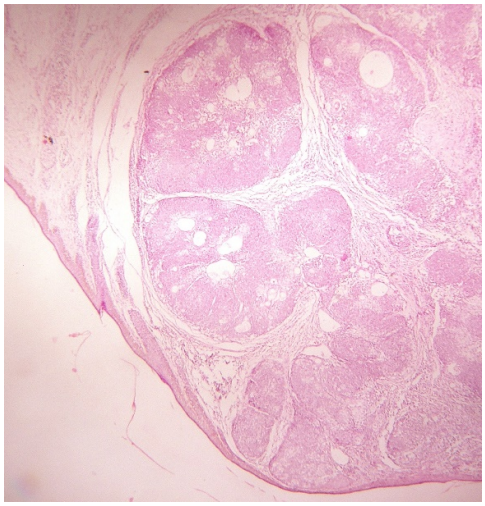

Figure3.H & E Nodular growth of biphasic cells

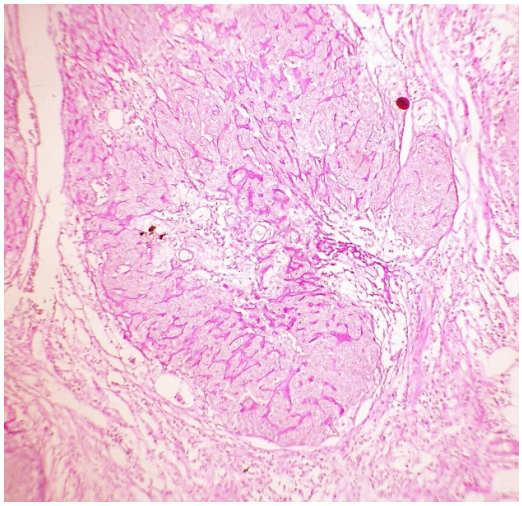

Figure 4 PAS Clear cells show PAS positivity.

Follow up of 2 years did not reveal any recurrence.

Solitary tumors of neural tissue origin are a rare occurrence inside the oral cavity. Neurofibroma are benign tumors that arise from peripheral nerve sheath. [5] Even rarer is the intrabony neurofibroma. This is because nerve sheaths or myelinated nerves are not present inside bone. [6]

Case Report

A 72-year-old male presented with a complaint of an intraoral soft tissue growth in relation to the lower right posterior teeth. Examination revealed a circular soft mass, 2.5cms in diameter, arising from attached gingiva and mucosa in relation to mandibular molars (Figure 5). An excisional biopsy was performed.

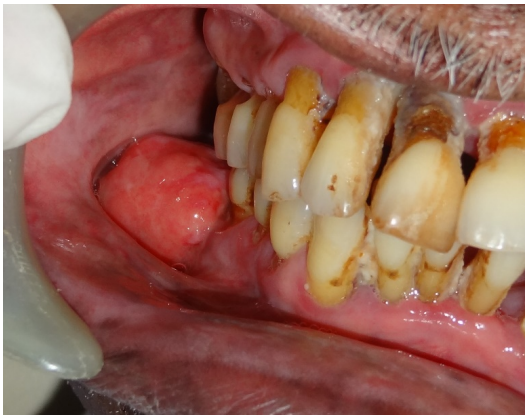

Figure 5 Lesion seen in right gingivobuccal sulcus

Histopathologic examination revealed parakeratinized stratified squamous epithelium overlying areas of myxoid to moderately collagenous connective tissue stroma. The stroma was highly cellular and exhibited interlacing nerve bundles with buckled nuclei. Mast cells and areas of calcifications were present. Chronic inflammatory cell infiltration of plasma cells and lymphocytes, blood vessels and Russell bodies were evident (figure 6,7).

These features were suggestive of a diagnosis of neurofibroma.

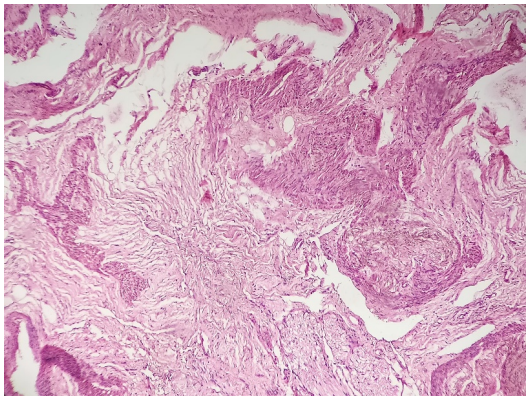

Figure 6 Nerve bundles in a background of a collagenous stroma

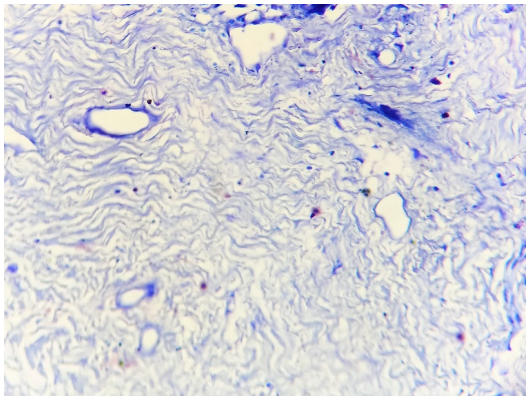

Figure 7 Toluidine blue mast cells

Case Report

A 66-year-old female presented with a chief complaint of pain and discomfort in the lower left premolar region of the jaw. Intraoral examination revealed edentulous residual alveolar ridges. No swelling, bony expansion or tenderness was present. There were no subjective or objective findings of paresthesia. A CBCT examination revealed a radiolucent ovoid lesion about 14mm x 10mm in close proximity to mental foramen. There was resorption of the buccal cortical plate (Figure 8). An excisional biopsy was performed. Intraoperatively, an intrabony soft tissue mass was found with evidence of infiltration into adjacent bone (Figure 9). The mass was not in direct continuity with the inferior alveolar nerve.

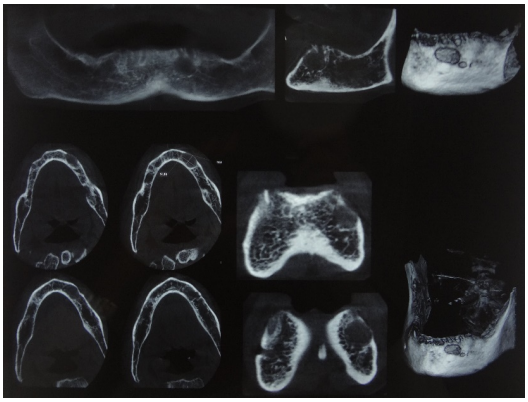

Figure 8 CBCT Note proximity to inferior alveolar canal

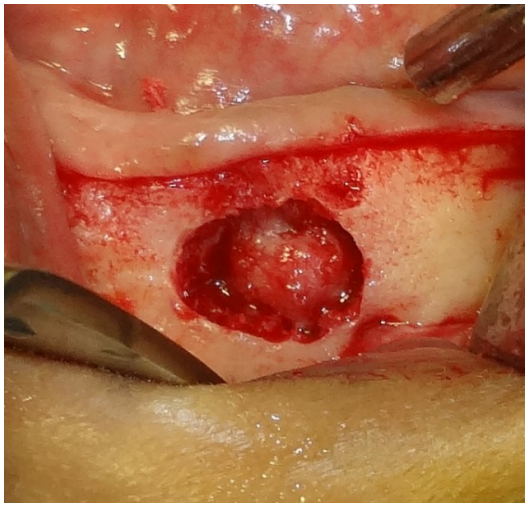

Figure 9 Intraosseous lesion. Note proximity to mental foramen

Histopathologic examination revealed an unencapsulated, well circumscribed lesion consisting bundles of spindle shaped cells with wavy nuclei. The stroma exhibited thick bundles of collagen intermixed with mast cells, chronic inflammatory cell

infiltrate, chiefly lymphocytes and plasma cells, focal areas of nerve tissue and cholesterol clefts (Figure 10). The lesion tissue was seen to invade associated bone. A histopathologic diagnosis of neurofibroma was made.

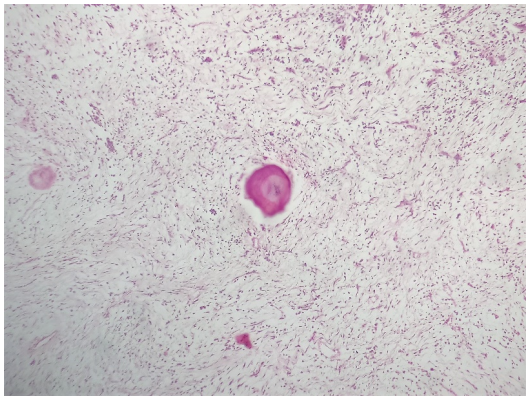

Figure 10 Myxoid content with spicules of calcification

In both of these cases careful follow up did not reveal any recurrence or other manifestations.

Low Grade Mucoepidermoid Carcinoma of Sublingual Salivary Gland

Sublingual salivary gland tumors account for 0.3-1% of epithelial salivary gland neoplasms. [7,8] Sublingual tumors are mostly malignant, Adenoid Cystic Carcinoma (ACC) and Mucoepidermoid Carcinoma (MEC) being the common histologic types. Clinically, they present in the floor of the mouth as an asymptomatic mass. [9] Treatment of choice is primarily surgical followed by radiotherapy for high-stage and grade tumors, close or positive margins. [10]

A case of low grade MEC of the sublingual salivary gland is presented.

Case Report

A 56-year-old female patient was referred for the management of an asymptomatic swelling in the left sublingual region of the anterior floor of the mouth. A low grade MEC of the left sublingual salivary gland had been diagnosed 4 years ago. A surgical excision of the tumour had been performed elsewhere. 4 years later a firm nodule in the region of the left sublingual salivary gland was again detected.

On examination, a 2.5cm X 1cm oval, firm swelling was present with no pain or mucosal ulceration (Fig. 11). Fine needle aspiration was performed. An ultrasound examination of the neck did not detect any neck nodes. The tumour was staged as T2N0M0.

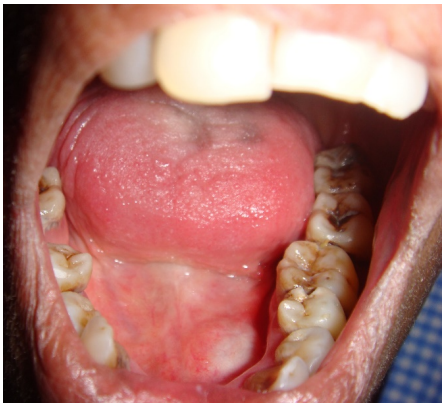

Figure 11 Swelling in left sublingual salivary gland region

Under general anesthesia using an intraoral approach, an en bloc resection of left sublingual salivary gland and tumour,

adjoining tissues of the floor of the mouth, mandibular alveolus and teeth was performed. The defect was closed with an inferiorly based nasolabial flap (Fig. 12).

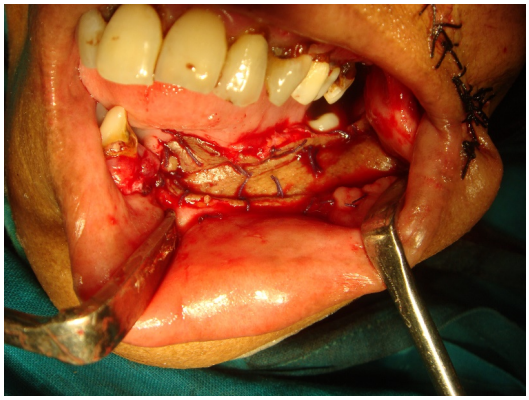

Figure 12 Resection and Reconstruction

Histopathological examination provided the definitive diagnosis of low grade MEC of the sublingual salivary gland (Figure 13,14). Resection margins were clear of tumour. The patient was placed on follow up and remained asymptomatic five years after surgery.

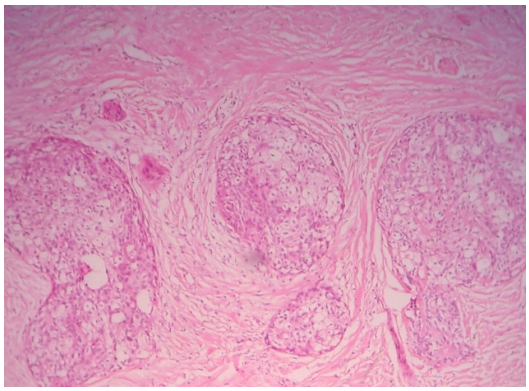

Figure13 H&E Low Grade Mucoepidermoid Carcinoma

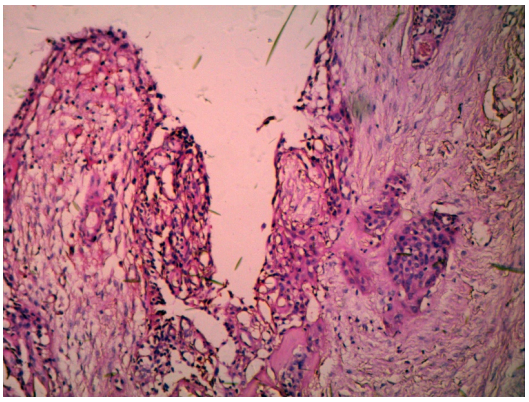

Figure 14 H&E - low grade MEC

Discussion

Hidradenoma is almost always benign and local recurrence is very rare. Occasionally, though, lymphatic invasion and lymph node metastasis has been reported in tumors which are histologically benign. [11]

Treatment suggested for this lesion is local excision. However, if the lesion is diagnosed to be hidradenocarcinoma, metastases can occur. [12,13] The malignant form does not show any gender or age predilection. Some of the criteria for malignancy include poor circumscription, nuclear atypia, deep extension and necrosis. [14] Malignant hidradenocarcinoma is very rare with aggressive clinical course and distal metastasis. [15]

Neurofibroma are benign tumors that may occur singly or as part of neurofibromatosis (Von Recklinghausen's Disease of the skin). Tongue is the most frequent location of intraoral neurofibroma. [16]. Intraosseous

neurofibroma is usually seen in posterior mandible and maxilla. [17] Both the intraoral and intraosseous lesion illustrated in this paper have occurred in uncommon locations. Whatever the location, neurofibroma tend to bleed during surgery due to their extensive vascularity and this should be kept in mind during surgical planning. [18] The tumors can be enucleated from the nerve trunk; resection of the nerve is not indicated. [19]

Solitary intraosseous neurofibroma may be a manifestation of neurofibromatosis. [20]. Recurrences and possibility of malignant changes make long term follow up essential. [21]

Mucoepidermoid carcinoma is rare, the 'Low Grade' malignancy is well differentiated whereas 'High grade' shows poor differentiation. Whatever the morphological sub-typing, radical resection of primary lesion has to performed. All mucoepidermoid carcinomas are poorly radiosensitive. [22] Elective neck dissection is recommended in high grade histology or large (>4cm) tumors. [23]

Conclusion

The lesions illustrated in this paper are distinct entities with differing tissues of origin. The common factor is the site of occurrence in the maxillofacial region, intraoral or extraoral. This is of clinical significance and, though rare, warrants their inclusion in differential diagnoses.

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